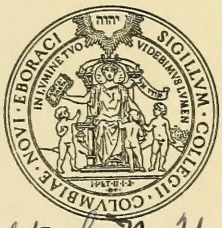

Base Hospital No. 20,

TEXT-BOOK

ON

NERVOUS DISEASES

BURR

TEXT-BOOK

ON

NERVOUS DISEASES

BY

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AUTHORIZED ENGLISH EDITION
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WITH 156 TEXT ILLUSTRATIONS

VOL. I

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THE·MAPLE·PRESS·YORK·PA

PREFACE TO THE AMERICAN EDITION

The German contributors to this book should need no introduction to American physicians as they are all men of established reputation. The articles are based on personal knowledge and experience and each writer presents his own well thought out and mature opinions. While the work is purely scientific in tone and conservative in attitude, a matter of much importance in these days when much wild doctrine, especially as to mental therapeutics, is being offered the public, it has never been forgotten by the writers that the only useful knowledge is that which leads to sane therapeutics. There is no need to discuss the advantages offered by a book written by many authors over a one-man work as it has been done by the editor of the German edition. The translators have endeavored to do their work faithfully and to give an accurate rendering of the writers' statements. The American editor has not cut out the many references to European health resorts, not because he thinks Americans need go abroad for such treatment, but in order to encourage Americans to develop and encourage their own natural cure places.

EDITOR OF THE AMERICAN EDITION.

PREFACE TO THE GERMAN EDITION

The marvellous advance made by neuro-therapeutics during the second half of the last century has, as a natural consequence, induced specialization within the confines of this domain. On the one hand this tendency was furthered by the unlike scientific descent of the leading investigators, among whom from the very first, there was a clear cut distinction between the psychiatrical- and the interno-neurologists; on the other hand, neurology, in itself, was a field to which not a few borderland fields naturally attached themselves.

Thus it has come to pass that the neuro-therapeutics of to-day stands in intimate relation not only with psychiatry and internal medicine, but also with ophthalmology and otology, with dermatology, bacterio-serology, and above all with surgery, making use more and more of the aid afforded by these special sciences.

If therefore we venture to submit to physicians and students a newly compiled text-book of neuro-therapeutics, we are not blindly following the fashion—a fashion which in recent years has given extraordinary precedence to this cooperative method of text-book production—but are conforming in no slight degree to that very peculiarity which characterizes, as we have said, our field. Our intention was to produce for students and practitioners —that is essentially for non-specialists—a text-book comprising old material, that has long been established, as well as the new, nay the very latest acquisitions of neuro-therapeutics. It aimed to give those who have but little time for specialization a book containing what is essential, concise, and vitally instructive, though naturally devoid of that fullness to be found in the special monographs. How could these requirements be better met than by having the separate chapters written by authors whose work and original successful researches prove that they stand in the very midst of the events and the literature of each field concerned? Within the narrow confines of a text-book, they are able to unite, first of all, brevity with clearness, and completeness with the necessary conciseness. A few examples may serve to illustrate: Clinical treatment of disturbances of the circulation of the brain, however much it concerns therapeutically the special neurologist, considering its close relation to the pathology of the heart and the circulation, doubtless demands presentation by an internist whose special subject is the pathologic physiology of the circulation. The discussion of meningitis, also, will be less adapted to the special neurologist than to the internist skilled in bacteriology and cyto-diagnosis. On the other hand, the exposi-

tion of the present condition of the pathologic physiology of the brain necessitates, in view of its complicated structure, and the rapid progress made in recent times, a specialist's treatment. It is no accident that this difficult realm is almost or quite a terra incognita to many interno-neurologists of the well known schools and that it is to a small degree only cultivated by many representatives of clinical psychiatry. And finally: of all the neurologists, internists and psychiatrists, who should undertake to present briefly and critically the modern status as to the operative possibilities in neurotherapeutics? Every one will here give precedence to the surgeon of this special field, and we are glad and proud to have found as collaborator for this special field the most noted German brain surgeon.

I think that I have shown by these few examples that the compiling of a text-book that recognizes the newly-acquired results, by *several* authors, may well be justified. That disadvantages may, to some extent, offset the advantages of this method, can not be denied, and corresponds to the experience with compiled text-books in other fields of medicine. On account of the multiplicity of authors, the uniformity of presentation necessarily suffers, nor can repetitions be altogether avoided. We ask the reader, therefore, to judge these faults leniently, and hope to obviate them to some extent in later editions.

Should we succeed in arousing by this text-book, in wider classes of non-specializing physicians, interest for and understanding of neurology as an equally justifiable and important special field among those other special fields long officially recognized as being of signal importance, we shall see in this success at once the attainment of our purpose and our reward.

THE EDITOR.

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GENERAL DIAGNOSTICS
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VOL. I

I

GENERAL DIAGNOSTICS OF NERVOUS DISEASES

BY

S. SCHOENBORN (Heidelberg)

1. TAKING THE HISTORY

The dictum that medical art in its clinical aspect is deprived of its best foundation when lacking a good history of the patient, sounds very old-fashioned to-day, in this age of orthodiagraphy and precipitin reactions. Should I assert, however, that this dictum is nevertheless correct and for no special field more correct than for nervous diseases, I should not like to be summoned, on this ground, as star witness for a supposed backwardness in neurology. On the contrary, I would state, that, rather it is an advantage that within the confines of our field that endless splitting up into extreme specialism, recent and very recent, has not as yet taken place, as we see it to-day in several branches of internal medicine. A summary of our knowledge of neurology, in spite of the wealth of new observations and facts increasing, too, year by year, is still possible for the individual physician; he need not yet, because of the confusing mass of separate sciences, lose a clear view of the case as a whole—that great inheritance of the first neurologists and internists of the last century. But for this he needs, over and above the modern equipment of diagnosis, before all, the crude tools of those masters: the history of the patient.

This history of the patient consists according to the traditional scheme of anamnesia, of status, diagnosis, prognosis and therapy. I find no reason for making any changes in this scheme. I know of no more comprehensive one. The method of "genius diagnoses" ("snap" diagnoses) at first sight imposes only on the novice. However certainly the experienced *may* recognize from a distance a tabetic by his staggering gait and the easily visible pupillary differences and a case of *Basedow's disease* by the staring eyes and the goiter, he must never *rely* on his empiricism. Very often exact anamnesia alone suffices to correct the would be cock-sureness of diagnosis at first sight.

I said that the value of an exact history of the patient is especially high, precisely in nervous diseases. This holds true, to begin with, of the *anamnesis*. It is more indispensable here than in most internal diseases, though

on the other hand, it is less often possible to make the diagnosis from the anamnesia alone, than in these diseases. A sufferer from kidney trouble may by an exact account of the development of the disease, and of his troubles alone, assist the physician to a correct diagnosis, and, on the other hand, in this case, the physician can usually make the diagnosis by the examination alone, even without any anamnesia. Both are but rarely possible with the nervous patient. He can not off-hand with sufficient exactitude so state his troubles with respect to time and place as to make the disease recognizable by them alone, especially since, in nervous diseases, a slowly progressing course with a large number of slighter and more severe symptoms is the rule. But almost without exception, not even the most minute examination, alone, can enlighten as to the genesis of the disease and the troubles of the patient, which are of such great therapeutic importance.

In taking up the previous history we inquire first about the family history. Are there among the ancestors or descendants, mental diseases, characteristic neuroses, alcoholism? Were the parents consanguineous? Did the parents suffer from syphilis probably or possibly? (Mercurial treatment, premature births.)

Nor can the unfortunately all too indefinite question about "nervousness in the family" be avoided, and it often brings to light important facts.

All these factors may be comprised under the designation "neuropathic taint."

Then follow the questions as to the previous diseases of the patient. Here, one must consider before all: normal birth; children's diseases, especially of the nervous kind (spasms, infantile convulsions, migraine, pavor nocturnus, enuresis, etc.), and the thereto predisposing disturbances in development (rachitis); infectious diseases (measles, scarlet fever, typhoid, diphtheria, influenza); chronic use or exposure to certain poisons (alcohol, nicotine, lead, morphine); over-exertions of every kind (mental, physical, especially too, sexual); masturbation. Particular attention, furthermore, is to be given to two points: first, the possibility of preceding syphilis, for which one should watch with the greatest care and the greatest objectivity (chancre, eruptions, mercurial treatment, symptoms of visceral syphilis, miscarriages and abortions in women; the vocation is to be considered also) secondly, accidents or injuries experienced, whereby, of course, in the interest of the patient, too clear a *reference* to the traumatism is, *at first*, to be especially avoided by the questioner. Furthermore, one must investigate the mental development (schooling) and note the vocation of the patient.

The *previous history and the history of the present trouble* conclude the anamnesis. While, in his former questions, the investigator may proceed according to a scheme, here, with minute consideration of all nervous regions, he must often deviate from the scheme and follow a definite idea,

which may lead him in the direction of fixed disease pictures; for, as has already been mentioned, the nervous patients scarcely ever succeed in sensibly arranging their symptoms in proper order and even the schematic questioner has before him, at the end, usually a chaos of individual symptoms, a disease history "without head or tail." In particular, the physician should attempt to ascertain the very *beginning* of the disease, its time and place; the correct diagnosis often depends on the temporal sequence of the symptoms (for instance spinal atrophies). Therefore, in suspicion of tabes one will proceed from possible eye symptoms, lancinating pains, in a myelitis, from paræsthesias in the feet, bladder disturbances, etc.

Naturally the symptoms in the internal organs (stomach, heart), must be considered also.

It should, as a rule, be possible to ascertain from the history all symptoms of a nervous disease, that *can* at all be subjectively perceived.

2. THE OBJECTIVE FINDINGS

A. General Remarks

In the examination following the history the mere observation of the nervous patient may, in itself, be of great value. Facial expression and the attitude of the body, especially in bed-ridden patients, may give much information. A rigid attitude or carriage of the body may be dependent upon spastic pareses (myelitis, system-diseases, central affections), paralysis agitans, upon meningitis, tetanus, and other affections. Pains in certain nervous regions often induce anomalies in the posture of the body, which mostly have in view a relaxation of the nerves concerned (sciatica), or cause involuntary twitchings of the musculature in these regions (tic douloureux). *Asymmetries*, too, strike the attention here at once, not only those that affect the musculature (cf. below), but also especially those which affect the structure of the bones. Scolioses and kypho-scolioses cause suspicion of the presence of sciatica, compression myelitis, syringomyelia. Asymmetries of the face may rest upon progressive facial hemiatrophy, sympathetic affections, acromegaly. In the latter case, we find similar asymmetries in the extremities. Cranial asymmetry may be due to tumors or hydrocephalus. Nor must we overlook *congenital malformations* (cranial malformation, abnormal ear formation, hare-lip, polydactylia, etc.), but one may say that this branch of neuropathic taint has more real importance in psychoses than in nervous diseases (probably most in the so-called neuroses).

There are rarely found in nervous troubles noticeable changes in the skin and the visible parts of the mucous membranes. Here, again, traces of syphilis must be most carefully followed (*Hutchinson's* triad, scars from tertiary processes, swollen glands). Circumscribed, more rarely diffused reddening or cyanosis, also anæmic pallor, may be interpreted as a sympathetic

disease, occurring alone, or with spinal or cerebral troubles, even with neuroses. Neurogenous edemas are found in hysteria, more rarely in spinal diseases. Ulcerative changes we see also in hysteria (mostly through self-inflicted injuries), likewise as severe trophic disturbances, usually combined with synchronous disturbances in sensibility, especially in tabes and syringomyelia, finally as decubital sores in every long continued loss of local sensation (also in the mucous membranes—keratitis neuroparalytica). Rare trophic neuroses sui generis (*Raynaud's* disease) may lead to ulceration and gangrenous destructive processes. The blister-like eruptions of herpes zoster belong to a dermatologic-neurologic borderland; they are combined nearly always with neuralgias in the nervous region concerned. We shall have to discuss all these points in more detail later.

The physical methods of internal medicine in general use are of but small service in the examination of nervous patients. *Percussion* may occasionally be employed on the skull, in internal affections of the cranium; a peculiar flat tinny percussion note (reminding of the bruit de pot fèlé), the so-called "*cracked pot note*" is occasionally found on the diseased side in cerebral tumors. Palpation sometimes serves for the recognition of anatomic changes in the peripheral nerves and nerve sheaths. It may also be important in discerning the causes of a dysbasia arteriosclerotica by the absence or the smallness or the hardness of the peripheral arterial pulse. *By auscultation*, we recognize in rare cases the presence of vascular tumors and aneurysms of the arteries inside the cranium. As an aside we may remark that auscultation and percussion naturally may be of use to us indirectly also in neurology, in the recognition of the causes of peripheral paralyses (recurrens paralysis in aortic aneurysms), etc., and that the establishment of *sensitiveness to pressure* in bones and nerve trunks (caries, neuralgias) may be of decisive importance.

Great weight must from the first be laid upon the correct interpretation of disturbances of consciousness and the psyche. Dimming of consciousness is, on the whole, easily recognized; the lightest form is usually designated *somnolence*, a condition of sleepiness from which the patient may however be awakened by a mere call, in which correct answers can be obtained, the next higher degree as *sopor*, with retained reflexes, in which awakening is produced only by means of violence (by stimulation of the skin, etc.), the highest degree as *coma*, with the reflexes lost. Here the patient can not be awakened even though force is used. All three forms occur in most cerebral diseases as well as in all those internal diseases that affect the central nervous system.

Knowledge of psychic disturbances in detail belongs to the realm of psychiatry, with the fundamentals of which, however, every neurologist must be acquainted. How important are even the lightest forms of these disturbances, often how typical! The patient's mood is occasionally charac-

teristic of his disease; the euphoric mood is seen mostly in multiple sclerosis, in *Friedreich's disease;* gloomy morose psychic states in paralysis agitans; exalted (maniacal) moods in progressive paralysis (which, as a fact, all but belongs to the purely psychic realm). Stupor, the almost reactionless depressive state rarely occurs in pure nervous diseases; more frequently the talkativeness of most neurasthenics which must not be at once assigned to the sphere of maniacal conditions. Especially typical is it in *traumatic neuroses*, in which, of course, the exclusion of the patient's desire for lucre and of conscious or unconscious simulation is one of the most difficult tasks of the neurologist.

In order to have a clear picture of the psyche of nervous sufferers, it is important to win their confidence, to allow them to talk.

Then gradually, by clever inquiries, the investigator may discover the severe psychic symptoms: delusions, illusions, hallucinations. Disturbances in memory, especially for *more recent* events, are common and important. Intelligence is often considerably clouded (dementia, imbecility, idiocy), sometimes congenital, with especial frequency the result of diseases during infancy (hydrocephalus, encephalitic processes) somewhat less frequently acquired at an advanced age (multiple sclerosis, epilepsy, progressive paralysis of the insane, etc.). For many disturbances in memory and intelligence, the ability to *count* is of peculiar importance; therefore one should set the patient problems with this in view. For all psychic conditions, finally, the *writing* of the patient may be significant, which furthermore discloses numerous disturbances in motility (ataxia, all forms of tremor, writer's cramp, etc.).

All of the lighter and some of the more severe psychic disturbances are found in the restricted field of neurology, if we except progressive paralysis with the enormous variety of its psychic symptomatic picture, in all the inflammatory processes inside the cranium, in cerebral tumors, hydrocephalus, traumatisms of all sorts, in multiple sclerosis, in some "system" diseases (*Friedreich's* ataxia), in that strange combination of a multiple neuritis with psychic disease, known as *Korsakow's* psychosis, and in numerous neuroses (neurasthenia, hysteria, epilepsy, chorea, paralysis agitans, etc.).

The general examination of nervous patients in respect to visible external changes, the results of physical examination in the narrower sense and of that of the psyche, are to be followed by the examination of the organs of sense.

B. Testing the Sense Organs, Voice and Speech

1. *The Eye*

Every test of the function of the *organ of vision* should begin with an examination of the *pupils.*

In the normal state, both pupils are of equal width; *congenital inequality* (anisocoria) occurs rarely and in contradistinction to other authors we do not interpret it as a sign of a neuropathic disposition. In rare cases, this inequality alternates between the two eyes; each eye in alternation with the other shows now a *miosis* (contraction) now a *mydriasis* (dilatation). We know as little of the origin of these "rapidly alternating" pupils as we do of the so-called *hippus*, the rapidly changing width of the pupil of one eye without external stimulation.

Usually, however, pupillary differences signify a disease of the tracts subserving this contraction and expansion, at least in one eye. Here, the wider as well as the narrower pupil may be the one diseased (no absolute values exist for pupillary width in any form that can be used by neurologists). To decide whether both eyes or which of the two is the one affected, we test the *pupillary movements.*

They consist in a *contraction* of the pupils controlled by the m. sphincter pupillæ (oculomotorius), which appears reflexly, when light falls upon the eye and upon accommodation, as voluntary concomitant movement, with convergence and energetic contraction of the orbicularis oculi, and in a *dilatation* of the pupils controlled by the m. dilator pupillæ (sympathetic), which appears reflexly in the dark, and upon pain stimulation (for theory and localization of pupillary movements see special section in this text-book).

Testing of the *reaction to light* is best carried out in a darkened room, by means of a reflector (forehead-, eye-mirror) throwing concentrated light, coming from a source behind the patient, into one eye. This is the best way of establishing the normal, rapid and extensive contraction of the pupil under examination, the absence of reaction, the *reflex-pupillary rigidity* to light (*Argyll-Robertson's* phenomenon) as well as all the intermediate steps, the most important of which is the "sluggish" reaction, a slow contraction of the pupil, comparable in its course to several other pathological reflexes (*Babinski* toe reflex).

Almost simultaneously with the process in the illuminated eye there appears in the non-illuminated, the so-called "consensual" contraction; observation of it may be important for the determination whether reflex disturbances are to be localized in the centripetal (opticus) or the centrifugal (oculomotorius) reflex tract, since in the latter case, the non-illuminated pupil contracts consensually in spite of the areflexia of the illuminated pupil, and not in the former.

Theoretically, sluggish reaction and loss of the reaction to light of the pupil may of course appear in every disease that interrupts the reflex tract at any point. Practically, however, this is the case only in relatively few well characterized diseases of the central nervous system, of which the metasyphilitic (tabes, progressive paralysis), meningitis and tumor cerebri may be mentioned as the most frequent. The rigidity to light may be uni-

lateral or bilateral; pupils with total rigidity to light are somewhat more frequently miotic, contracted (often to the size of a "pin head"), than mydriatic, dilated.

Naturally, testing the effect of light falling upon the eye may—and this will be the rule in investigations as to orientation—take place in daylight; one then has the patient (this is the best way) look through a well lighted window at a distant object and fixate it, and covers—this is to be recommended in all cases—both eyes at the same time, and then by taking away the covering (hand, etc.), tests separately the direct and consensual light reaction of each eye. But this test may be made exceedingly difficult by poor daylight, very dark iris, or by the light reflected from the cornea.

Independently of the pupillary contraction upon entrance of light, *contraction upon accommodation* or *convergence* may take place. Normally, both pupils energetically contract upon accommodation of the eye for near objects (if the mm. interni are normal, with simultaneous convergence). This contraction may be retained in the absence of the light reaction (frequently in tabes) or it may be missing at the same time as the light reflex (tabes, progressive paralysis, cerebral tumors) constituting total pupillary rigidity (ophthalmoplegia interna); a case of isolated accommodation paralysis occurs very rarely and is practically negligible.

The voluntary contraction of the pupil upon innervation of the orbicularis oculi is easily tested, but has not yet attained pathological importance.

Reflex dilatation of the pupils is far less important. We test it by shading the eye in day light, as well as by pain stimulations (pinching or pricking the skin of the cheeks). The undubitably justifiable conclusion that an absence of the dilatation reflex, must, in general, point to an affection of the sympathetic, can be used but rarely neurologically with our slight knowledge of the processes in the sympathetic region. On the other hand, we find this absence frequently with the synchronous absence of contractions to light in the pupils, in tabes, progressive paralysis of the insane, severe affections of the substance of the brain; the pupils in this case are neither contracted nor dilated to a high degree, but are of medium width.

Abnormal miosis may depend upon absence of the reaction to light as well as upon paralysis of the dilator pupillæ, and also (rarely, almost only in hysteria) upon a sphincter spasm. In analogous manner, we see mydriasis in sphincter paralysis, spasm in the dilator pupillæ and not infrequently as a purely psychic reaction in terror, great mental exertions, etc. Neurasthenics often have permanently very dilated pupils.

Finally it must be mentioned that in old age all the pupillary reflexes usually decrease, and that certain poisons tend to dilate the pupil (atropin, hyoscin), others to contract it (eserin, morphine). In chloroform narcosis, the pupils, as a rule, are immobile to light and dilated.

Next to the test of pupillary reaction, comes that of the *external muscles*

of the eye. Here are concerned the m. rectus ext. (n. abducens), obliquus sup. (n. trochlearis), obliquus inf., rectus int., levator palpebræ (all the n. oculomotorius), while the orbicularis oculi, supplied by the facialis, is to be dealt with in the test of musculature of facial expression.

Besides the action of the *m. levator palp.*, which raises the upper eyelid, only the *m. rectus ext.* and the *rectus int.* are to be tested in simple manner, by asking the patient to look energetically towards the right or the left, or better, with head fixed, to follow with the eyes, as long as possible, an object that is moved in the direction to the right or to the left. If one of these two muscles is hindered from acting, the eye concerned does not follow in the direction of its normal contraction. A second necessary consequence is the appearance of *double images* (with retained vision), which, however, are frequently consciously or unconsciously suppressed by the patient, since he fixates with one eye only. In slight pareses of one muscle, the test for double images is often the more delicate as contrasted with that for eye movement; for this purpose one asks the patient to say when the object which is being slowly moved sidewards is seen by him double or confused, and in the former case, in what relation the double images stand to each other. To facilitate the recognition of both images (of which one is usually seen more brightly, that is, more clearly), one may place a red glass before one eye; the red and the white image can then, even though partly overlapping, be easily distinguished from each other. In paresis of a rectus int., the double images are at the same height and are "crossed," i. e., the image that "belongs" to the *right* eye lies on the *left* side; in paresis of the rectus ext. the double images are at the same height and "uncrossed."

Examination of the obliqui and the rectus sup. and inf. is more difficult, since their effects are always somewhat combined so as to move the eye upwards, downwards, or in a circle. The *rectus inf.* moves the eyeball downwards and inwards, the *obliquus sup.* downwards and outwards and rotates it somewhat; the *obliquus inf.* upwards and outwards, the *rectus sup.* upwards and inwards, with synchronous rotation. In these muscles, the loss of movement is, as a rule, very slight and difficult to test; it is best to examine the double images, for which the following scheme holds good:

In paralysis of the rectus inferior, the double images appear only on looking down, the images are crossed, inclined to each other, and one below the other, the false image is the lower, and upon lowering of the object concerned, separates from the true image, which is higher.

In paralysis of the obliquus superior, double images appear upon looking up; the pictures are uncrossed, inclined, and one below the other.

In paralysis of the obliquus inferior, double images appear upon looking up; they are inclined, one below the other, but uncrossed.

In paralysis of the rectus superior, double images appear upon looking up; they are inclined, one above the other, and are crossed; the false upper

image leaves the true lower image upon raising of the object used in the test.

Weakness of the eye muscles causes still other phenomena of functional loss, which, however, are of less diagnostic service to the neurologist. These are for every single muscle the *secondary contracture* of its *antagonist*, which, when vision is turned directly forwards, conditions an abnormal position, a *strabismus* (most clearly in paresis of the rectus int.—strabismus divergens, and in paresis of the rectus ext.—strabismus convergens); furthermore, occasionally, the *secondary deviation* of the unaffected eye, the abnormal position of the head, and finally, upon an attempt to exert the paralyzed muscle, some twitchings of the bulbus in the sense of the working of this muscle (so-called paresis-nystagmus, especially in conditions of weakness in convalescents).

Naturally besides the paralysis of individual, and the paralyses of all the external muscles (ophthalmoplegia externa), or of all the eye muscles (total ophthalmoplegia), there occur also variously grouped paralytic conditions in several muscles. Of these should be mentioned the so-called *conjugate paralysis of the eye muscles*, with loss of the power to turn both eyes laterally in the same direction (affecting, therefore, only one rectus int. and ext. of each eye); here the convergence of bulbi is usually retained in spite of the necessary co-operation of the paralyzed internus concerned. Then there is *convergence paralysis*, the counterpart of the disturbance just mentioned, in which both bulbi retain their ability to turn laterally, and then the rare divergence paralysis, which is difficult to recognize (the patient can not bring back to their normal position the bulbi which have been converged to look at an object near by).

Heretofore we have spoken only of the paralyses of the individual muscles. The *muscular groups* supplied by one of the *cerebral nerves* mentioned, may also be paralyzed on one or both sides. When the *n. abducens* and the *n. trochlearis* are paralyzed, the disturbances may readily be deduced from what has already been said. In paralysis of the *oculomotorius*, the eye affected is, as a rule, fixated towards the outside downwards; and of active movements, there is possible only a slight outward rotation of the bulbus. Furthermore, in total oculomotorius paralysis, the pupil concerned is dilated, and, as a result of paralysis of the levator palpebræ, the upper eyelid droops (*ptosis*). This ptosis may appear also as an isolated phenomenon (in hysteria, occasionally likewise in organic diseases as for instance tabes). For the rest, there occur isolated and grouped paralyses of the eye muscles in a great many central and peripheral (eye-muscle-nerve) affections of the central nervous system, among which must be mentioned above all, the syphilitic and metasyphilitic diseases (tabes, progressive paralysis) as well as some toxic-infectious processes (diphtheria).

As we shall see later, in all disturbances of motility in the human body,

a second group of disturbances besides paralysis may be thought of, i. e., hyperkinesis, *spasms*, in which we distinguish *tonic* (continuous tension of the diseased muscle) and *clonic* spasms (change between tension and relaxation, twitching movement). This holds also for the eye muscles, in which, however, spasms are very much rarer than paralyses. Thus a strabismus based upon genuine spasm (usually convergent) is very rare, observed almost exclusively in hysteria. Let us mention here that by far the greatest number of all cases of strabismus (so-called muscular or concomitant squint) are not based upon a muscular paralysis, but, so to say, only upon conditions of weakness, a so-called disturbance in the equilibrium of the muscles; that in spite of the changed position of the bulbi, it proceeds usually without double vision and disturbances in motility, and that as a result, the typical test for the individual eye muscles gives a negative result. Cf. on this point text-books on ophthalmology. We must sometimes regard as tonic spasm, the remarkable phenomenon of *conjugate deviation*, in which both eyes are held fixed in a position of rotation towards the right or the left (especially in cerebral tumor in certain regions, often accompanied with rotation of the head to the same side). In rarer cases, it may rest also upon a paralysis of the antagonists.

The various forms of *nystagmus* may be interpreted as *clonic spasms* of the eye muscle: short twitchings following one another in slow or rapid succession, of one or both bulbi in a lateral direction (nystagmus horizontalis), in a vertical direction (N. verticalis), or in rotation (N. rotatorius). We find it (cf. above)frequently in pareses of individual muscles in respect to and in the direction of their activity (especially N. horizontalis, which, moreover, in "*end positions*" *of the bulbi*, in the extreme lateral direction, are not infrequently found in the healthy, too), furthermore, congenital (here often very intensive, when vision is directed straight forward), as vocational disease (miners), in blindness and finally—this is of especial importance for the neurologist—in a series of affections of the brain, above all in multiple (cerebro-spinal) sclerosis.

While testing the individual eye muscles is one of the most difficult chapters in the diagnostics of nervous diseases and now and then makes assistance from an ophthalmologist desirable, examination of the *background of the eye* should, in its main points, be well mastered by the neurologist. We may exclude here the real affections of the retina and chorioidea, which less frequently occupy the attention of the neurologist (at most occasionally as sign of neuropathic disposition, as in retinitis pigmentosa and other congenital diseases), but must consider that sometimes the diagnosis of a cerebral disease seems to be erroneously confirmed by an affection of the retina, as in a retinitis albuminurica confined to the disc and simulating a neuritis optica.

Two ophthalmoscopic findings especially are of interest to us:

Neuritis optica, or *choked disc* and *atrophy of the optic nerve.*

In neuritis optica the disc appears clouded, reddened, or grayish red in color, the veins are dilated, the arteries of the disc contracted, the edge of the disc not clearly marked. The diameter may be considerably increased. In pronounced choked disc, there is, besides, an ophthalmoscopic, usually easily recognizable protrusion of the disc; the blood vessels appear to be snapped off at the edge of the disc, and disappear in the diffuse redness of the enlarged disc.

Concerning differentiation between neuritis optica (papillitis) and choked disc, it is all the more difficult, considering what has been said, inasmuch as there is by no means a consensus of opinion as to the cause of the disease. A mechanical congestion (cerebral tumor) through increase in the intracranial pressure (accumulation of large quantities of cerebro-spinal fluid inside the cranium) and a gathering of fluid in the sheath of the optic nerve may give rise to "papillitis" as well as choked disc; on the other hand, toxic products in infectious and other diseases may evoke choked disc also in addition to the neuritis optica, or papillitis which usually results. In general, the ophthalmologist will incline rather to a differentiation of the two conditions, the neurologist to the identification of them, with the restriction that they both represent *different degrees* of the same affection.

The acuteness of vision in neuritis optica is nearly always normal in the beginning of the disease, and very frequently later also, at least central vision, whereas contractions of the visual field (cf. below) frequently appear later; but the beginner must never forget that there may exist even a high degree of choked disc without the knowledge of the patient. Thus the ophthalmoscopic picture here by no means corresponds to the degree of the visual disturbance.

The recognition of choked disc is in nearly all cases easy; only the lightest degrees of incipient papillitis may cause difficulties; of course one must not be deceived by the apparent indistinctness of the edges of the disc in refraction anomalies.

Naturally we find choked disc most frequently in processes which cause congestion, especially in tumor of the brain. Here it is usually (but by no means always) bilateral. The situation and size of the tumor are not always decisive as to the origin and intensity of choked disc, still, very large tumors can scarcely run their course without accompanying papillitis, while, on the other hand, we must remember that frequently in recent times there have been observed cases of typical tumor symptoms with choked disc, though in the operation or the autopsy, no tumor was found (Nonne). Meningitis of any kind may cause papillitis; it is found with special frequency in meningitis serosa and in hydrocephalus. In all forms of cerebro-spinal syphilis, in lead poisoning, in acute infectious diseases, in chlorosis, it occasionally occurs; a rheumatic inflammation of the opticus, comparable to

"rheumatic" paralysis of the eye muscles is also observed. On the other hand, it is rarely or never found in encephalitis, nor in apoplexies of any kind, and it is lacking almost without exception in two diseases which are accompanied by very typical changes in the background of the eye, that is, in tabes and in multiple sclerosis. In both cases, as a rule, we find in its stead atrophy of the optic nerves.

Atrophy of the optic nerve is ophthalmoscopically recognizable by the abnormal pallor of the disc, which may lead to a porcelain white coloration or discoloration, but which may also present only a slightly paler color in one-half of the disc. In the latter case, diagnosis is nearly always difficult, since, as is well known, there are present in normal conditions differences in clearness in the separate parts of the disc, and especially the part first affected, the temporal half of the disc is mostly subject to these fluctuations. In these incipient cases, there is lacking, too, that sharp almost hard outline of the white disc against the red retina, that is so characteristic of the final stage. The width of the vessels is usually not changed to any considerable extent.

Atrophy of the optic nerve may appear as a primary phenomenon, and it may also represent the final stage of a neuritis optica, that is, be a secondary phenomenon. The latter, which may occur in all the diseases mentioned above, is usually accompanied by considerable disturbance in vision (especially loss in the visual field), whereas primary atrophy may exist for a long time without any subjective disturbance, and is therefore often overlooked (sometimes, especially in multiple sclerosis, there occur also sudden transitory visual disturbances of high degree, though the background of the eye shows nothing else but the always equally slight "temporal paling" of the disc). Except when it follows traumatism, opticus atrophy is nearly always bilateral, though often developed in different degree in the two eyes. Primary atrophy of the optic nerves is found almost only in *tabes*, progressive paralysis of the insane, and *multiple sclerosis;* in the former case it is usually total and progressive; in the latter partial and more or less stationary. Whether as a matter of fact, it is not frequently secondary in multiple sclerosis, arising after a neuritis belonging to the prodromal stage so often mistaken for a "hysteria," must, owing to the scarce findings from examination at this prodromal stage, remain undecided.

Cases of so-called *neuritis retrobulbaris* occur rarely in the practice of the neurologist, but more frequently in that of the ophthalmologist. Ophthalmoscopically, they are recognizable not at all, or by an (secondary) atrophy of the papilla of moderate degree; on the other hand, they show the typical visual disturbance of a central loss in the visual field for colors (red and green). Retrobulbar neuritis is usually toxic, or toxic-infectious, and occurs as an isolated phenomenon or with a polyneuritis which owes its origin to the same causes. The most common causes are chronic poisoning

with alcohol or nicotine, also, diphtheria. A medicinal neuritis retrobulbaris occurs also. Frequently retrobulbar neuritis optica has been observed as a very early prodromal symptom of multiple sclerosis (*Uhthoff*, and others).

The real examination of *visual disturbances* in nervous diseases—test of acuteness of vision—is, in the main, the task of the oculist. But nevertheless, it is evident that no neurologist will fail to attempt an approximate determination with the assistance of *Snellen's types*, of counting fingers at a certain distance, etc. An examination of the *visual field*, which is generally easily feasible, is earnestly recommended. While one covers one of the patient's eyes, and asks him to fixate a point with the other, at the distance of from one-half to one meter, small pieces of white or colored paper are introduced into the visual field area, and one moves the pieces of paper from the outer edge of the visual field (that is, behind or above or near the patient's head) slowly in the direction of the point fixed by the patient up to that point itself, and then tells the patient to say when he catches sight of them first and whether they disappear again on the way (central scotoma). For practical reasons, one chooses as the main direction of these movements, first the main meridians, above and below, outside and inside, then eventually an intermediate meridian. All this can be done far more easily with the various perimeters, some of which are quite cheap, on which one can read the degrees directly (degrees in width of the approximately half spherical field of vision). One must consider that the visual field in normal condition is variously extended (for white above, 50° to 60°; below, 60° to 70°; outside, 90°; inside, 60°; for colors, considerably less) and that individual differences occur also. One must never, especially when using a perimeter, fail to enter the results upon printed diagrams of the visual field, as this is the only way to get an accurate picture of the visual field.

Disturbances in the visual field occur with and without other visual disturbances, and with or without changes in the background of the eye. We differentiate the concentric contraction, the peripheral scotoma, the central scotoma and hemianopsia. The *concentric contraction*, a diminution on all sides (for white and colors, or only for colors) of the visual field, as well as *scotoma, the loss in spots*, which, according to the location of the spot in the visual field, is designated *central* (near the point of fixation) or *peripheral*, we find, as a rule, in neuritis optica (papillitis or neuritis retrobulbaris) or in atrophy of the optic nerve. In contraction, the disease of the nerves is more frequently diffuse, in scotoma it strikes only some bundles of fibres. Therefore we find both disturbances in tabes, multiple sclerosis, polyneuritis, in the processes above mentioned which lead to choked disc, and, finally, isolated also; the concentric contraction is frequent too in hysteria.

Hemianopsia, the loss of an entire half of the visual field is less frequent. Here, it is a question nearly always of the loss of the outer or inner (not the upper or lower) half, and, moreover, preponderantly in both eyes at once,

corresponding to the partial crossing of the fibres of the optic nerves. One speaks of *homonymous* (bilateral) hemianopsia, if both right or both left halves are lost; on the other hand, of bitemporal hemianopsia (*heteronymous hemianopsia*), if both external halves of the visual field are missing. A binasal hemianopsia occurs virtually never. Hemianopsia is easily recognized by the test suggested for the visual field.

Hemianopsia is found in lesions of the chiasma and of the tractus opticus; and, moreover, the latter produces homonymous hemianopsia, and, on the other hand, the lesion of the chiasma (at least of a middle portion containing the crossing of the fibres of the optic nerve) leads to bitemporal hemianopsia. Diseases of the chiasma are, as a rule, evoked by small tumors at the base of the brain, while the tractus may naturally be affected by every agent that injures the substance of the brain. Especially important are the cases in which a homonymous bilateral hemianopsia presents the only sign of a lesion (hemorrhage, tumor) of the occipital lobe, in which the visual center is situated.

Hemianopsia, in contrast to concentric contraction, is very clearly present in the consciousness of the patient, since it is frequently combined with other disturbances, concentric contraction of the field of vision, visual hallucinations, etc. Theoretically, moreover, every hemianopsia belonging to the tractus opticus should exhibit an objective symptom; there should be no pupillary reflex to light thrown upon the eye, if one illuminates only the "blind" half of the retina (hemianopic pupillary rigidity). This does occur occasionally, but is often difficult to demonstrate.

Monocular diplopia, double vision in one eye alone, an abnormality occurring almost exclusively in hysteria, does not belong to the real organically conditioned disturbances of the apparatus of vision; nor does *exophthalmus*, the protrusion of one or both eyes. We find this phenomenon unilaterally in tumors of the orbit or—through congestion—also in tumors of the interior of the cranium; bilaterally, above all, in Basedow's disease. In Basedow's disease the origin of the symptom is not altogether clear; but it probably depends upon a swelling of the retrobulbar tissue, such as occurs, and sometimes periodically under other conditions.

2. *Hearing*

While examination of the eyes has proved to be very useful, strangely enough, the value of the *ear findings* has so far proved to be comparatively slight for neurological diagnosis. In part this is due to the fact that genuine nervous diseases of the ear are really rare; in part also to the difficulty of diagnosis in these affections. Only very recently have clinicians (*Wittmaack*) begun to look with more interest upon the diseases of the inner ear.

Three groups of symptoms are of use in neurologic—otiatric diagnostics;

subjective auditory disturbances (difficulty in hearing, hyperacusis, tinnitus aurium, etc.), objective changes in the ability to hear (*Weber's* and *Rinne's* experiments) vertigo, nystagmus, disturbances in equilibrium. All the other resources of the otologist, especially the whole of otoscopy, may at most be used indirectly by the neurologist, for the entire group of disturbances in the sound conducting apparatus does not allow any conclusions to be drawn, as to the presence of a nervous disease, excepting only anomalies, like the occasional occurrence of laxness of the ear drum through atrophy of the m. tensor tympani in advanced dystrophy.

Of the *subjective auditory disturbances*, it is most difficult to use tinnitus aurium, ringing or buzzing of the ears, etc., for local diagnosis. This occurs, it is true, especially in affections of the inner ear, which, however, may be of most varying types: arterio-sclerosis, neuritis acustica, transitory disturbances, usually of unknown origin, such as tinnitus aurium of chlorotics, the ringing in the ears which appears paroxysmally in *Ménière's* disease, etc. Hyperacusis (oxyecoia) is found in paralysis of the facial nerve (cf. above), and in some neuroses, especially in hysteria. Every degree of difficulty in hearing occurs in diseases of the inner ear, in those that are really "nervous" (labyrinth and cochlea) as well as in those of the sound conducting apparatus (tube, tympanic cavity, drum, auditory meatus), and does not, upon superficial examination permit of differential diagnosis between these two groups. This first test of the ability to hear is made as follows: after telling the patient to close one of his ears, words or numbers are whispered or he is allowed to hear the ticking of a watch, and the limit of audition is measured in meters (from the ear). *Rinne's* and *Weber's* experiments afford a more delicate test, in as much as both are concerned with the *conductivity of the bones of the head.*

(*a*) **Rinne's Experiment.**—The handle of a tuning fork pitched at low c, after being set in vibration is placed upon the mastoid process of the affected ear. In normal cases, when the sound is no longer audible to the patient by this sort of "bone conduction" perception is still possible through "air conduction," if one holds the fork close to the ear (Rinne's positive test"). The case is *the same*—usually with diminished intensity of perception—if there be present difficulty in hearing of nervous, labyrinthine type. If, on the other hand, Rinne's test is "negative," that is, if the sound be heard longer by means of the bones than through conduction by air, which occurs only in difficulty in hearing—this defect must be traced back to the sound conducting apparatus.

(*b*) **Weber's Experiment.**—A vibrating tuning fork is placed upon the middle of the frontal or occipital aspect of the head. In normal conditions the sound is heard by each ear with equal intensity. Should, however, a *unilateral* disturbance in hearing obtain—and the experiment is valuable only in such conditions—and the sound be more perceptible by the better

hearing ear, then the disturbance lies in the *reception* (nervous) apparatus of the other, namely the affected ear. But should the patient localize the sound in the less perceptive ear then the disturbance lies in the sound *conduction* apparatus of the affected ear.

We must mention, furthermore, that in diseases of the inner ear, as a rule, the distinctness of audition is impaired especially in regard to the *higher* tones.

Disturbances in equilibrium, nystagmus and **vertigo,** in so far as they proceed from the ear, are probably always to be traced back to the inner ear, the nervous apparatus (also indirectly in diseases of the tympanic cavity). The former occur spontaneously especially in the peculiar attacks that were formerly supposed to be characteristic of the general concept: "*Ménière's* disease," and besides this, may easily be evoked in pathological cases (galvanization with weak currents). A feeling of dizziness (ear vertigo), occurring in attacks as well as continuously, is very rarely lacking in disease of the inner ear. A real nystagmus is, as a rule, evoked in labyrinthine affections, upon turning the axis of the body. According to the most recent views, these symptoms also make possible a differential diagnosis between disease of the n. vestibularis, or of the labyrinth, and of the n. cochlearis, or of the cochlea and the organ of *Corti;* to the n. vestibularis belong disturbance in equilibrium and the feeling of dizziness, to the cochlearis, the nervous auditory disturbance, in a stricter sense.

"Nervous" auditory disturbances are found in fracture of the base of the skull, tumors of the base, arterio-sclerosis, in the changes due to old age, and known as "otosclerosis," in multiple sclerosis, in tabes dorsalis (as genuine degeneration of the nerves, comparable to atrophy of the optic nerve), in isolated neuritis, or in neuritis complicated with polyneuritis (*Frankl-Hochwart*), hysteria, neurasthenia, and especially in traumatic neurasthenia (the favorite hypothesis of "labyrinthine concussion").

3. *Taste and Smell*

The *sense of smell* is of least neurological usefulness, in spite of its so frequent disturbances in daily life; disorders are mostly dependent upon catarrhs of the mucous membranes, not upon diseases of the olfactory nerves themselves. Above all, therefore, one must establish in neurological investigation whether the olfactory pathways are clear, and no catarrh is present. If everything is well in both these respects, one must test the acuteness of the sense of smell (naturally for each nostril separately) by holding odorous substances in front of the nose, avoiding of course those which stimulate the sensory end organs of the trigeminus (ammonia). As a rule, I use asafœtida, tincture of valerian, oil of cloves and oil of roses (or eau de Cologne). A gradation of the acuteness of the sense of smell is all the less

possible for the various odors, since the delicacy of the sense of smell, the "good nose," varies exceedingly in normal conditions and in a large number of healthy persons is developed very scantily. The odorous substances mentioned, should, as a rule, be recognized.

In hysteria there occur *hyperosmia* (hyper-sensitiveness) and *anosmia* (lack in sensitiveness to olfactory stimuli). In tumors of the base of the cranium, of the ethmoid bone, etc., we find occasionally unilateral lowering of the ability to smell, likewise, in meningitis and fractures.

Somewhat more useful is the test of the sense of *taste*. A prior condition, however, is that the sense of smell be not simultaneously disturbed (catarrh in the head). We conduct the test as follows: with a little glass rod we place in succession a drop each of solutions representing the four main qualities of taste, sweet, sour, salt, bitter—sugar, vinegar, salt, quinine—on the outstretched tongue in this manner:

(a) on the anterior half of the tongue, separately on the right and left sides;

(b) on the posterior half of the tongue, separately on the right and left sides.

After applying each drop of the solution one waits a few seconds, then wipes off the tongue energetically with a dry cloth, has the patient withdraw his tongue and state the taste concerned. We do this for the following reasons: In respect to the sense of taste, the anterior half of the tongue is under the dominion of the chorda tympani, that is, of the n. lingualis; the posterior half and the palate under that of the n. glossopharyngeus. Therefore the test is made separately. The tongue is drawn back because many patients have no taste sensations upon the *extended* tongue. In general sour is better recognized on the tip of the tongue, bitter on the posterior half of the tongue.

Disturbances in taste come to be diagnostically important almost only in diseases of the n. facialis for clearer localization (chorda tympani), also, however, in disturbances of the n. trigeminus III. The diseases of the glossopharyngeus which are rare in themselves, are concerned only exceptionally. Progressive paralysis and neurasthenia (especially their depressive forms) are frequently accompanied by disturbances in taste, which in that case, however, affect the entire tongue.

4. *Disturbances in Voice and Speech*

These processes belonging to the realm of the cerebral nerves, if not to that of the actual nerves of sense, may be briefly discussed here.

Disturbances in *voice* occur mainly in two forms: as voicelessness (*aphonia*) and as *hoarseness;* abnormally high pitch of the voice (squeaking voice, etc.), is rare, occurring almost exclusively in hysteria. Aphonia also, in the

great majority of cases in which it comes to the notice of the neurologist, is of hysteric, that is, of psychogenous, origin, but even detection of other stigmata, etc., must not, as occasionally happens, deter the investigator from laryngoscopic examination. For even if it occurs rather frequently that a hysteric aphonia is treated for weeks as chronic laryngitis, still, a patient suffering from catarrh has sometimes been branded as hysteric—the worse

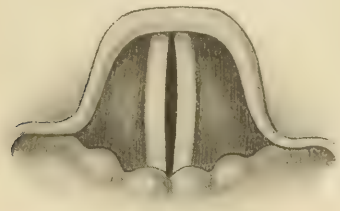


FIG. 1.—Bilateral paralysis of the posticus in the moment of inspiration. (*After Ziemssen.*)

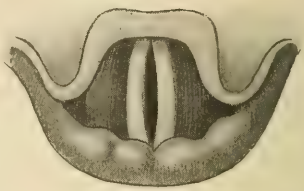


FIG. 2.—Paralysis of both N. thyreoarytænoidei interni. (*After Ziemssen.*)

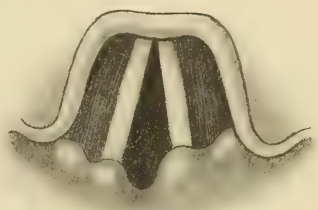


FIG. 3.—Paralysis of the recurrens at the left side (position during inspiration). (*After Ziemssen.*)

FIG. 4.—Paralysis of the arytænoideus. (*After Ziemssen.*)

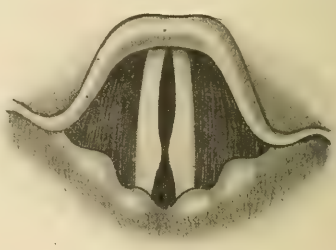


FIG. 5.—Bilateral paralysis of the thyreoarytænoidei with synchronous paresis of the arytænoideus. (*After Ziemssen.*)

fate! The laryngoscopic examination must therefore be as familiar to the neurologist as the ophthalmoscopic. The technique and the details of the laryngoscopic findings do not belong here, but the disturbances first mentioned (aphonia, hoarseness) are nearly always the consequences of a lesion of the n. laryngeus inf. (recurrens), which according to its severity, presents very different laryngoscopic pictures. Of these we shall mention the three most important.

1. *In total recurrens paralysis*, the vocal cord concerned (or in bilateral paralysis, both vocal cords) remains during phonation and inspiration in the same immovable position between the ad- and abduction (cadaver position).

2. In so-called *internus paralysis* (paralysis of the thyreo-arytænoidei interni) the vocal cords are not altogether closed in phonation, but leave an oval slit open; they remain lax, and consequently hoarseness results.

3. In so-called *posticus paralysis* (paralysis of the cricoarytænoidei postici) phonation is as a rule normal, but there usually appears inspiratory dyspnœa, since the muscles mentioned are dilators of the glottis, and therefore upon their loss, the vocal cords are brought pathologically nearer to each other, are drawn in. In unilateral paralysis there is only inspiratory stridor; bilateral posticus paresis may, especially upon exertion and the resulting forced breathing, lead to severe dyspnœa (Figs. 1 to 5).

All these conditions—there are in addition a number of rarer laryngeal pareses, all of which cannot be mentioned here—are found in recurrens, or in vagus affections, which localized peripherally more frequently than centrally, may be evoked through tumors, traumatisms, aortic aneurysms, etc. Hysteria may simulate each of these pictures, so that it may be difficult to distinguish an hysterical from an organically conditioned (nervous) aphonia. Organic recurrens paralyses are more frequently unilateral, hysterical usually bilateral; but one must remember that also in chronic catarrhs slight pareses of the type described may occur.

Of the *disturbances in speech*, the most important group, the aphonic symptom complex belongs to a special chapter (cf. its discussion in a special part of the volume). We are concerned here only with the disturbances in *articulation* ("speech utterance disturbance" "Sprachstoerung," *Schuster*) which are usually designated as "anarthria" or "dysarthria." It is a question here exclusively of innervation disturbances of the muscles needed in speech (lips, tongue, palate, also larynx and respiration) or of the nerves subserving them (facialis, hypoglossus, vago-accessorius, phrenicus) and in many dysarthric disturbances one can recognize at once from the act of speech which group of muscles is attacked when the patient is speaking. When all the muscles of the tongue, palate and lips are affected, there results only an unintelligible babbling, total anarthria, as we usually find it in the final stage of bulbar paralysis. But this disease usually shows an affection of the separate muscles progressing gradually in groups, and these too are not easily differentiated. In paralysis of the tongue, e and a, also d, l, r, s, t, are spoken indistinctly; paralysis of the lips makes the utterance of, oo, o, b, f, p, v, difficult or impossible. Paralysis of the muscles of the soft palate makes speech nasal (since part of the air expired passes through the nose in phonation), and separate letters (g, k, German ch) also become indistinct.

Stuttering is dependent upon another disturbance. Here it is a matter, partly of a tonic, partly of a clonic spasm in the muscles used in speech, and, moreover, not only the lips and tongue, but also the muscles of respiration are affected at the same time. Frequently it is not the spasmodic tension of a certain muscular group that takes away from the patient the possibility of getting beyond the initial letter (which most often causes the trouble) but the *synchronous* tension of *various* muscles, working partly as antagonists. In contrast to anarthria, stuttering must be looked upon as a functional neurosis, and is scarcely ever dependent upon an organic disease.

By "*scanning*" we designate a speech disturbance in which the single syllables follow one another slowly, rhythmically, at clearly defined intervals. It is (with rare exceptions in hysteria) an organically conditioned central innervation disturbance, which is characteristic of multiple sclerosis, and finds its parallel in the spastic rigidity of the muscles of the extremities which occurs in this disease.

The *stumbling over syllables* in patients suffering from progressive paralysis, represents a mixture of anarthria on a motor and on a psychic basis. In the utterance of long words—favorite words for the test are "Massachusetts artillery," "truly rural"—the first syllables are spoken correctly, the following ones stumblingly, or are mutilated or partly omitted. Obviously there is concerned here in addition to the mere disturbance in articulation, a forgetting of the separate syllables during speech, or failure to understand the word. Frequently the paralytic does not notice his speech disturbance, in contrast to other persons suffering from dysarthria, who are keenly conscious of their defect.

Besides the disease forms already mentioned, we find anarthria or dysarthria, in *peripheral* nerve affections (hypoglossus), occasionally congenital, and commonly in all sorts of diseases of the cortex of the cerebrum (tumors, apoplexies); in these latter, frequently with a genuine aphasia, especially in recent apoplexies, which must not be overlooked when testing.

Mutism, the total inability to speak, is congenital, except for its appearance in hysteria and will be discussed in that chapter.

5. *Disturbances of the other Cerebral Nerves*

belong here only in the minority of cases, since the motor nerves (facialis, trigeminus III, accessorius, hypoglossus) will be mentioned when discussing motility, the sensory trigeminus, when discussing the sensibility test, with the symptoms of which the forms of disease, in the main, coincide. But we recommend to the physician who is making a systematic examination always to institute an investigation of all the cerebral nerves preceding that of the peripheral nervous system. In addition, we shall mention here only the disturbances affecting the soft palate, and the complicated mechanism representing deglutition.

The *soft palate* (mainly innervated by the vago-accessorius group) participates in speaking, swallowing (and in opening of the Eustachian tube). We find corresponding symptoms of disturbance: 1. In phonation (one has the patient pronounce ah) the diseased half of the palate is not raised, but hangs down laxly, the soft palate and the uvula are in a position slanting towards the sound side (the uvula even in normal persons is not always quite straight). 2. Upon swallowing, food (especially liquid food) passes to the nose because of the insufficient closure of the pharynx. 3. Occasionally, slight auditory disturbance. Paralyses of the palate are found most frequently in polyneuritis (diphtheria), then in central affections (bulbar paralysis).

In *deglutition* the muscles of the lips, tongue, palate, pharynx and œsophagus are involved. Consequently, different disturbances in swallowing appear, according as to whether only one and which one of these groups is affected. In weakness of the lips, liquid flows out of the mouth, in weakness of the tongue, it cannot be transmitted to the pharynx, or the food remains in the cavities of the cheeks.

Since all swallowing of liquids is effected by means of pressure of the tongue against the hard palate, patients with paralysis of the tongue are often unable to take any liquid nourishment at all; paralysis of the palate causes, as has been said, part of the food to pass into the nose. Finally, a paralysis of the muscles of the pharynx and œsophagus causes only a disturbance in the swallowing of solid food, which, in such cases, is pushed down only with great trouble, that is, sinks down of itself. Unilateral deglutition paralysis gives rise to almost the same disturbances and produces the feeling of "food going down the wrong way," which is caused by entrance of part of the food into the larynx and its reflex reaction.

Disturbances in deglutition are found most frequently in toxic and infectious polyneuritis and in central nervous diseases (bulbar paralysis, etc.).

Other symptoms of the glosso-pharyngeal-vagus group will be mentioned in the special sections devoted to neuritis of the cerebral nerves.

We may remark here in anticipation that *electric changes in the cerebral nerves* may be established with certainty only in the facialis, trigeminus III, accessorius, hypoglossus, and the soft palate.

C. Motility

The condition and activity of the musculature represents the first of the three main groups in the examination of the nervous system (especially of the peripheral).

Inspection of the musculature can distinguish, above all, three things—*hypertrophy*, *atrophy*, and, to a certain degree, the *tonus* of the muscles. To ascertain the presence of hypertrophy or atrophy, especially the pathological

degrees of both, one naturally needs an exact knowledge of the normal forms of the body, that is, of the normal (or nearly normal) forms of the muscles. Now there is scarcely any other tissue of the body which, through exercise or lack of exercise or through weakening diseases, etc., undergoes greater changes than the musculature. The great muscles of the pugilists may bear the most striking external similarity to the pseudo-hypertrophic muscles of the sufferer from dystrophy, and the patient with immobilized joints may, in respect to the appearance of his muscles very easily be confused at first sight with a case of severe polyneuritis. A less practised eye than is concerned with the above changes is, however, somewhat more useful, if it be a question of recognition of differences in the musculature between the right and the left side, between peripheral and central parts. But here, too, the eye alone must not decide, though doubtless it best recognizes slight differences. One must employ an exact *measurement* likewise, which, in so far as musculature alone is concerned, usually signifies only measuring the circumferences of the extremities. Naturally, for measuring, exact symmetrical points must be chosen (measuring the distance from fixed points upon the bones in centimeters), and it must be considered furthermore that in right handed persons the right arm and the right leg have a circumference of about one to two cm. more than the corresponding extremities upon the left side. The trained eye can naturally detect easily symmetrical atrophies, especially those in the extremities (hands), but to establish these changes *definitely*, one needs also an exact motility and electricity test. For tonus of the muscles cf. below.

Palpation of the muscles except for the rare determination of nodular thickenings (tumors, pseudo-hypertrophy) or a general softening of the muscular substance, affords no special conclusions:

All the more important is the *motility test.* In order to control this, one naturally needs an exact knowledge of the function of the individual muscles, which shall be here recapitulated briefly, in so far as it is concerned with testing the active movements. (I follow in the main *Strümpell's* division.)

1. **Mimetic musculature** (nervus facialis).

M. frontalis.

Function: Wrinkles the forehead into transverse folds.

Test: To look directly upwards. To draw up the eyebrows.

Loss in paralysis: After the order to draw up the eyebrows, absence of wrinkling on the diseased side.

M. occipitalis: Involved in the wrinkling of the forehead and the scalp.

Condition in loss and test the same as above.

M. corrugator supercilii.

Function: Wrinkles the forehead perpendicularly.

Test: Wrinkle your forehead!

Loss in paralysis: In unilateral paralysis, an often indistinct absence of wrinkles during the test.

M. orbicularis oculi.

Function: Eye closure.

Test: Close your eyes!

Loss in paralysis: The palpebral fissure can not be completely closed. Lagophthalmos.

M. compressor nasi, levator alæ nasi, zygomaticus, risorius, levator labii sup.

Function: Elevating the alæ nasi and the corner of the mouth.

Test: Turn up your nose!

Functional loss in paralysis: Corresponding; above all, loss of the naso-labial fold.

M. orbicularis oris.

Function: Pursing the lips, whistling.

Test: Corresponding command.

Functional loss in paralysis: Disability or inability to move the lips.

Atrophy of the mimetic muscles leads to sinking in of the parts involved.

2. **Chewing (masticatory) musculature.**

Mm. masseter and temporalis (N. Tringeminus III).

Function: Movements of chewing.

Test: Close your teeth tightly!

Functional loss in paralysis: Noticeable gap in the place of both muscles on the diseased side. Especially distinct in simultaneous atrophy.

3. **Movement of the soft palate; deglutition**: cf. above, under "other cerebral nerves."

4. **Muscles of the tongue** (N. hypoglossus).

Function: All independent movements of the tongue.

Test: Stick out your tongue, move it to the right, to the left, roll it upwards, downwards!

Functional loss in paralysis: In unilateral paralysis, the tip of the outstretched tongue deviates towards the *paralyzed* side (as a result of the peculiar radial distribution of the m. genioglossus). The movements of the tongue are all more or less hindered, but an isolated test of the other muscles of the tongue (m. lingualis, m. transversus linguæ) is of no great importance. In bilateral paralysis (bulbar paralysis, etc.), the tongue lies motionless upon the floor of the buccal cavity. *Atrophy* of the musculature of the tongue leads to the quite characteristic picture of the furrowed tongue, which shows uni- or bilateral transverse folds, irregular elevations and depressions and feels abnormally soft.

5. **M. sterno-cleido-mastoideus** (N. accessorius).

Function: Rotates and partially moves the head forward.

Test: Press the chin hard upon the fist of the examiner.

Functional loss in paralysis: The head is incompletely rotated, and bent crookedly forwards (the chin turns towards the paralyzed side). In the test described the characteristic cord of the muscle appears only on the sound side.

6. **Muscles of the Back.**

Mm. splenii, biventer, recti capitis postici (cervical nerves 1–4).

Function: Backward movement of the head and the cervical vertebræ.

Test: Lay the head backwards upon the supporting hand of the examiner.

Functional loss in paralysis: Oblique and incomplete performance of this movement.

7. **Mm. sacrolumbalis, longissimus dorsi, spinalis dorsi** (spinal nerves).

Function: Erects the spinal column.

Test: Rise from a stooping position against the resisting hand of the examiner, without help from the hands. (But cf. under 34.)

Functional loss in paralysis: Hindrance or inability to rise in the test described. In unilateral paralysis dorsolumbar scoliosis (often with lordosis) with convexity towards the paralyzed side. In bilateral paralysis: severe lumbar lordosis in walking (gait of dystrophics); in sitting often (not always) kyphosis.

8. **Abdominal muscles (recti, obliqui, transversus abd.;** dorsal nerves from the eighth on).

Function: Abdominal pressure; raising the body from a supine position. Flexing the spinal column forwards.

Test: Exert pressure as if evacuating the bowels. Rise from a supine position without help from the hands.

Functional loss in paralysis: Impossibility of rising as described. Difficulty in evacuation of the feces and urine. Lumbar lordosis, pot-belly. In unilateral paralysis, occasionally the linea alba and navel are drawn towards the sound side.

9. **M. quadratus lumborum** (plexus cruralis).

Function: Lateral movement of the spinal column.

Test: Corresponding to the function.

Functional loss in paralysis: Corresponding, generally unimportant.

10. **Diaphragm** (N. phrenicus from cervic. 4).

Function: Lengthening of the thoracic cavity in inspiration.

Test: Breathe deeply.

Functional loss in paralysis: Absence of the epigastric protuberance, retraction of the intestines and the diaphragm phenomenon (visible descending shadow upon lateral illumination) in inspiration; dyspnœa.

11. **Muscles of the shoulder girdle and arm.**

M. cucullaris or trapezius (N. accessorius).

Function: Elevates the shoulder blade and brings it nearer to the middle line.

Test: Raise your shoulder blades ("shrug your shoulders")!

Functional loss in paralysis: Since frequently only one of the three portions of the muscle is paralyzed, the picture may vary. The characteristic of cucullaris paralysis, restriction in raising the shoulders, belongs actually to the middle portions alone, whereas the upper (clavicular) only when the shoulder is fixated, draws the head somewhat towards the back, and the lowest causes the shoulder to move towards the spinal column. The typical picture of total paralysis of the muscles is: sinking of the acromion, lowering and forward position of the shoulder (and the arm), more or less pronounced horizontal position of the collar bone, and a moving away of the scapula from the spinal column. The *atrophy* of the muscle, which is usually present, leads to a flattening of the line between the shoulder and the neck and to a clearer prominence of the contours of the scapula (above all of the spine of that bone). This position of the shoulder blade is designated as "angel-winged."

12. **M. levator anguli scapulæ** (1 to 3 cervicalis).

Function: Elevates the inner superior angle of the scapula.

Test: Raise your shoulders. (Usually, if the cucullaris is retained, not easily recognizable); when the cucullaris is paralyzed, partly replaces it functionally.

Functional loss in paralysis: Slight (in so far as it is isolated). In combination with cucullaris paralysis; inability to raise the scapula.

13. **Mm. rhomboidei** (4 to 5 cervicalis).

Function: To bring the scapula nearer to the spinal column (especially the inferior angle).

Test: Draw your shoulders together in the back!

Functional loss in paralysis: The inner border of the shoulder blade stands out from the thorax (visible almost only when simultaneous cucullaris paralysis exists).

14. **M. serratus anticus major** (N. thoracicus longus, from cervical 5).

Function: Rotates the scapula about the sagittal axis, and fixates it when the upper arm is raised vertically; serves for fixation of the scapula to the thorax, even in a state of rest.

Test: Raise your arm above the horizontal position. Press your arm forwards against some resistance (pushing).

Functional loss in paralysis: In rest, frequently, a deviated position of the inner margin of the scapula, which, below, draws nearer the spinal column, at the same time rising a little from the thorax. The arm can no longer be raised above the horizontal line because of the insufficient fixation of the scapula. In pushing the arm forwards, the inner margin of the scapula moves forward like a wing from the thorax: "winged shoulder." The *atrophy* that is usually present at the same time allows the winged position

to become more clearly visible, and brings about a recognition of the usually easily seen serratus digitations in the lateral wall of the thorax.

15. **M. deltoideus** (N. axillaris).

Function: Elevates the arm to somewhat above the horizontal line.

Test: Raise your arm to a horizontal position (eventually against resistance).

Functional loss in paralysis: The arm can not be raised. The usually simultaneous atrophy is characterized by loss of the shoulder curve and by prominence of acromion and caput humeri.

16. **M. pectoralis major and minor** (N. thoracici ant. from cervical 5 and 6).

Function: Draws the arm up to the thorax.

Test: Press your outstretched arm against some resistance (hands of the examiner).

Functional loss in paralysis: Usually only weakness in the before mentioned adduction movement, since deltoideus and teres major afford partial compensation. In severe atrophy, the upper ribs may stand out prominently.

17. **M. latissimus dorsi** (N. subscapularis from cervicalis 5 and 6).

Function: Draws the upper arm backward and downward.

Test: With horizontally raised upper arm, press downward and backward upon the supporting hand of the examiner.

Functional loss in paralysis: Weakness in the test described.

18. **Mm. supraspinatus, infraspinatus, teres minor** (N. suprascapularis).

Function: Rotates the arm outward.

Test: Rotate the outstretched arm outward (against resistance).

Functional loss in paralysis: Weakness in the movement described. Sometimes disturbance in writing. The frequently simultaneous atrophy may be recognized by the flattening of the posterior arch of the shoulder blade.

19. **M. subscapularis, teres major** (N. subscapularis).

Function: Rotates the arm inwards.

Test: Rotate the extended arm inwards (against resistance).

Functional loss in paralysis: Weakness of the movement described.

20. **Mm. biceps, brachialis internus** (N. musculocutaneous).

Function: Flexes the forearm towards the upper arm without any rotation of the lower arm (slight supination).

Test: Flex your forearm when in a *supine* position (against resistance).

Functional loss in paralysis: Impossibility of flexing in this position; there appears at once pronation of the forearm. *Atrophy* of these muscles is easily recognized by the loss of the biceps curvature; the circumference of the upper arm is decreased perceptibly.

21. **M. supinator longus** (N. radialis).

Function: Flexes the forearm towards the upper arm in half *pronated* position (the middle position between pronation and supination).

Test: Flex your forearm against resistance with half pronated hand (palms towards the inside).

Functional loss in paralysis: In attempts at movement the hand is at once supinated. *Atrophy* can be easily recognized by a conspicuous gap in the musculature on the radial side of the bend of the elbow.

22. **M. triceps** (N. radialis).

Function: Extends the forearm.

Test: Extend your flexed arm against resistance.

Functional loss in paralysis: Impossibility of performing this movement against resistance.

23. **Mm. extensores carpi** (rad. and uln., N. radialis).

Function: Extends (that is, overextends), the hand towards the forearm.

Test: Extend your hand against resistance (at the metacarpus).

Functional loss in paralysis: Impossibility of performing the movement described; the hand hangs down limply from the extended, pronated, forearm.

24. **Mm. ext. digit. comm., indicator, ext. digiti v** (N. radialis).

Function: Extends the basal phalanges of the second to fifth fingers.

Test: Extend your fingers against resistance (at the basal phalanges).

Functional loss in paralysis: Impossibility of performing the movement described. The atrophy of all the muscles mentioned under 23 and 24 causes usually a moderate degree of wasting in the dorsal aspect of the forearm.

25. **Mm. interossei ext., and int., Mm. lumbricales** (N. ulnaris, also medianus).

Function: These groups of muscles abduct and adduct the fingers (only the interossei) at the same time, flex the basal phalanges, and extend the end phalanges of the fingers.

Test: Spread and press together against resistance the fingers that have been *extended* (it is best to do this against the fingers of the examiner which are inserted as in "folding the hands"). Flex the fingers against resistance (at the basal phalanges) and extend them against resistance (at the end phalanges).

Functional loss in paralysis: Impossibility of performing the movements described; above all, inability to hold on to thin objects (needle, coins) in the fingers. In a condition of rest, the 4th and 5th fingers deviate towards the ulnar side. There is no strength in pressure of the hand. *Atrophy* is made very typical by the sinking in of the spatia interossea and by the hyperextension of the basal phalanges resulting from the preponderance of the antagonists (the long extensors and flexors) with simultaneous flexion of the end phalanges. Wasting of the palm. The type of hand known as "claw hand" (paralysis of the ulnaris).

26. **M. palmaris, Mm. flexores carpi** (rad. and uln., N. medianus and n. ulnaris).

Function: Flexes the hand towards the forearm.

Test: Flex your hand against resistance (at the metacarpus).

Functional loss in paralysis: Usually slight; weakness of the movement mentioned.

27. **Mm. flexores digit. long. subl. and profund** (N. medianus).

Function: Flex the middle (sublimis) and end phalanges (profundus).

Test: Pressure of the hand with co-operation of the lumbricales and interossei. Eventually, isolated test of the profundus: pressing the tips of the fingers against tips of the flexed fingers of the inverted hand of the examiner, with attempts to flex the fingers (interlocking).

Functional loss in paralysis: Weakness of hand pressure. One must observe that simultaneous weakness of the extensores carpi also simulates weak hand pressure, since when the hand hangs down, the flexors mentioned are unable to functionate. Therefore with simultaneous radialis paralysis, the test can be made only with passively fixed wrist. *Atrophy* of 26 and 27 combined, cause wasting of the forearm (usually of moderate degree).

28. **M. flexor brevis and abductor digiti minimi** (N. ulnaris).

Function: Flexes the basal phalanx of the little finger and abducts the little finger.

Test: Flex your little finger against resistance (hooked in the finger of the examiner).

Functional loss in paralysis: Weakness of the movement mentioned. Atrophy of these muscles mainly causes the characteristic flattening of the ball of the little finger in paralysis of the ulnaris.

29. **Mm. extensor pollicis brevis and longus, M. abductor pollicis** (N. radialis).

Function: Extension, that is, abduction of the thumb and the metacarpus.

Test: Extend and abduct the flexed thumb against resistance.

Functional loss in paralysis: Weakness of the movement described. The thumb falls into the palm of the hand.

30. **M. adductor pollicis** (N. ulnaris).

Function: Adducts the thumb (metacarpus) towards the forefinger.

Test: Press your thumb against resistance towards the fixed forefinger.

Functional loss in paralysis: Weakness of the movement mentioned.

31. **M. opponens, abductor brevis and flexor brevis pollicis** (N. medianus).

Function: Flexion and opposition of the metacarpus 1 and the thumb (1 phalanx).

Test: Press your hand.

Functional loss in paralysis: Weakness in hand pressure. Often disturbances in writing. The *atrophy*, which is usually present at the same time,

flattens the ball of the thumb in characteristic fashion, and gradually brings the thumb into the direction of the other fingers, in the plane of which it finally lies: "ape hand" of the medianus paralysis.

32. **Muscles of the leg.**

M. ileopsoas (N. cruralis), *M. tensor fasciæ latæ* (1 and 2 lumbalis).

Function: Elevates the leg at the hip joint.

Test: Raise your extended leg from a supine position (against resistance).

Functional loss in paralysis: Disturbance in walking; walking is usually quite impossible, as is also raising the extended leg from a supine position.

33. **M. sartorius** (N. cruralis).

Function: Test like that under 32. Not infrequently, in paralysis of the flexors of the thigh and of the extensors of the leg proper the sartorius alone is spared.

34. **Mm. glutaei** (sacral nerves).

Function: Extension and slight abduction of the leg at the hip joint.

Test: In a supine position, press down against resistance the raised thigh. When standing, rise from a stooping position against resistance. Press the thigh outwards against resistance.

Functional loss in paralysis: Difficulty in rising from a sitting posture, in mounting stairs, in rising from a bent position (in common with 7), waddling gait.

35. **Mm. adductor brevis, longus, magnus, pectineus, gracilis** (N. obturatorius).

Function: Adduction of the thigh at the hip joint.

Test: Press the thighs together against resistance.

Functional loss in paralysis: Loss of the movement mentioned. Insecure seat when riding horseback.

36. **M. extensor cruris quadriceps** (N. cruralis).

Function: Extends the leg proper at the knee joint.

Test: In a supine position, extend the leg against resistance, while the arm of the examiner placed below the knee fixates the thigh in a slightly flexed position.

Functional loss in paralysis: Impossibility of extending the leg proper in the manner described. Severe disturbance in walking, though walking is possible with the leg in extension, as long as the patient seemingly supports himself upon the surface of the knee joint and does not innervate the flexors of the leg proper. Mounting stairs is, as a rule, impossible.

37. **M. biceps, semitendinosus, semimembranosus** (N. ischiadicus).

Function: Flexion of the leg proper and extension of the hip joint.

Test: In a supine position, draw up the leg (against resistance) towards the thigh.

Functional loss in paralysis: Inability to perform the movement mentioned, moderate disturbance in gait, inability to jump and run.

38. **M. tibialis anticus** (N. peroneus).

Function: Elevates the inner margin of the foot and extends the foot (subjects it to dorsal flexion).

Test: Scarcely possible as an isolated movement.

Functional loss in paralysis: Inconsiderable in isolated paralysis. The *atrophy* effects a characteristic sinking in of the muscular layer directly next to the margin of the tibia. Frequently the tendon of the overexerted ext. hallucis stands out prominently; the large toe is flexed dorsally.

39. **M. peroneus longus and brevis** (N. peroneus).

Function: Elevates the outer margin of the foot and subjects the foot to slight flexion (plantar flexion).

Test: As an isolated movement only possible electrically.

Functional loss in paralysis: In attempting extension, adduction of the foot appears; the great toe drags slightly upon the ground in walking; flat-foot develops.

40. **Mm. extensor digit. comm. longus, ext. hallucis longus** (N. peroneus).

Function: Elevates the toes and thereby the tip of the foot.

Test: Draw up the toes against resistance.

Functional loss in paralysis: The toes hang down limply. The participation of these muscles effects, mainly, however, in combination with 38 and 39, the picture of the peroneus paralysis: the tip of the foot hangs limply down, usually its inner margin is the lowest; as a result, it always drags upon the ground in walking. In order to avoid this the patient raises his thigh excessively when walking, the characteristic "steppage gait" results. Through secondary contracture of the antagonists (calf) there frequently develops pes equinus, or pes equino-varus. *Atrophy* of the anterior aspect of the leg proper.

41. **M. triceps suræ** (M. gastrocnemius, plantaris, soleus—N. tibialis).

Function: Plantar flexion of the foot.

Test: Press the foot downwards against the hand of the examiner; stand upon the tips of the toes.

Functional loss in paralysis: Loss of flexion of the foot. Inability to stand upon the tips of the toes or to dance. There frequently develops a pes calcaneus. *Atrophy* of this muscular group causes the most severe atrophy possible in the leg proper.

42. **M. flexor dig. comm. longus, brevis, flexor hallucis longus and brevis** (N. tibialis).

Function: Plantar flexion of the toes.

Test: Flex the toes downwards against resistance.

Functional loss in paralysis: Inconsiderable.

43. **Mm. interossei** (N. tibialis).

Function: Flexion of the basal phalanges.

Test: Scarcely possible in isolation.

Functional loss in paralysis: Inconsiderable. But through the atrophy of these muscles and secondary contractures of the antagonists, atrophy of the planta pedis and a "claw-foot" may develop.

Functional testing conducted in this way will always give a complete picture of the power of the individual muscles. For separate groups of muscles, functional power may be measured directly by means of the so-called *dynamometer* (*Charrière, Duchenne, Sternberg*), which, however, has been extensively used only for pressure of the hands. Here it gives good comparative values in respect to the strength of both hands and affords a comparison of the strength in various stages of the same disease.

The *degree* of the paralyses is important for their recognition. While total *paralyses* even of individual muscles alone will scarcely escape the notice of the examiner, mere conditions of weakness, *pareses*, are often very difficult to recognize; they are most easily recognized in unilateral paralyses through comparison with the same muscle on the other side. Naturally disturbances of the sensorium cause special difficulties in the recognition of palsies; here sometimes the tonus (cf. below) is of decisive importance; in recent apoplexies, the raised arm, even in stuporous patients, falls down far more laxly on the paralyzed side, as if dead, in contrast to that on the sound side. In small children, the determination even of complete paralyses is often difficult. Where it is not a matter of the more easily recognized disturbances in walking, the reaction of the child to pain stimulations often leads to the goal; one pinches or pricks the skin approximately in the region of the antagonists of those groups of muscles that one wishes to test; the degree of reaction to escape the irritation will usually determine whether or not weakness is present.

As to the *type* of paralysis, naturally nothing has been ascertained by the mere determination of its presence. We must emphasize here, that almost every muscular paralysis may be of *organic* or "*functional*" (psychogenous, usually hysterical) origin; but even among the organic paralyses, there is scarcely any point in the entire nerve-muscular apparatus which may not give rise to muscular paralyses: disease of the muscle itself (myositis, possibly dystrophy and myotonia), of the peripheral nerve (neuritides), of the spinal cord (diseases of the columns of the spinal cord, poliomyelitis, etc.), of the medulla, oblongata and brain (bulbar paralyses, apoplexies, etc.). The *paralysis* may furthermore, as we shall see below, be *flaccid* or *spastic* (with diminution or increase of the tonus), may be accompanied by atrophy or hypertrophy, etc. As to what form is concerned in the individual case, frequently electrical testing, sometimes only observation of the entire disease picture may decide.

In our discussion of the individual muscles, we have always noted when the *atrophy* of a muscle produces *characteristic changes in form.* Now this

happens more frequently in groups of muscles than in individual muscles; their decrease in volume produces characteristic pictures, which, for the experienced physician, make possible diagnosis at first sight. The pictures are often typical of certain well-determined diseases. We mentioned the "*claw-hand*" of ulnar paralysis, the "*ape-hand*" of median paralysis (of high degree), dorsal flexion of the large toe in paralysis of the tibialis anticus, etc. These pictures are found most frequently, especially in isolated forms, in peripheral neuritis, somewhat less frequently (and scarcely ever alone) in poliomyelitis, syringomyelia, bulbar paralysis. For progressive neurotic muscular atrophy, the peripheral beginning of the atrophy is characteristic; the extremities acquire that peculiar attenuation towards the periphery, which is known as "*bird-legs*." On the other hand, progressive spinal muscular atrophy also attacks the periphery first, but the atrophy here is of much higher degree. In syringomyelia we see that, as a rule, only the upper extremities show muscular atrophy; for poliomyelitis the apparently total participation of all possible muscular groups here and there, all over the entire body is characteristic. The most various and most easily remembered pictures are presented probably by dystrophy; the thin upper arms with the strong lower arm, the winged shoulders, lumbar lordosis, if the face is affected the tapir-lip, lagophthalmos. The number of such characteristic pictures may be easily increased, especially if one counts in all the changes in form dependent upon contractures (cf. below) with or without synchronous atrophies. For the sake of simplicity, we may here mention at once some other pictures of *disturbances in gait*, not directly connected with muscular paralysis and atrophy. We have just mentioned the "*steppage*" *gait*, the *waddling gait*, posture and gait in *dystrophy*. These kinds of gait belong to the *simple paretic* variety. All gait disturbances accompanied by lesion of the pyramidal tracts are to be designated as *spastic*, or *spastic paretic*. Common to them are spasms or tensions (cf. under hypertonia); in walking the legs are pushed forwards as if they were made of wood, flexed but slightly at the hip and not at all at the knee, the tips of the feet graze the ground, the soles of the shoes are always worn through at a definite place. Such is the gait in spastic spinal paralysis, lateral sclerosis, myelitis, mostly too in syringomyelia and multiple sclerosis. The *ataxic*, "*swinging*" or long-striding gait of the tabetics, the "*tottering*" gait of cerebellar ataxia, are mentioned below under ataxia. Persons suffering from *paralysis agitans* walk stiffly, mostly with bent back and bowed head, taking small steps, also "stick" to the ground, and still the gait is not really spastic; nay, rather upon occasion very quick tripping steps are made forwards and backwards, irresistibly, especially when with a light push, one sends the patient forward or backward or tells him to run. Then he cannot arrest his movements (symptom of *pro- and retropulsion*), which never occurs in spastics. Total inability to walk, known as "*abasia*" ("astasia"), which is not rare in hysteria, will be mentioned in the special part of the book.

The manifold picture of *writing disturbances* of a motor nature furnish a parallel to a certain extent to the *disturbances in gait.* Mere *paretic* disturbances, such as appear in isolated median or ulnar paralysis are but little characteristic; the patients write slowly, with trouble, but the writing is not characteristically changed, and occasionally retains wholly its previous character. Clumsiness naturally increases in complicated pareses (spinal muscular atrophy, multiple neuritis). There may appear total motor inability to write (*motor* agraphia). In *spastic* conditions the writing as a rule becomes smaller, labored, the separate letters run into one another, and the words often are illegible. *Ataxic* script (tabes superior) furnishes the type of deviating movements on paper; the letters are of varying length, single strokes pass across the letters already written, the lines are crooked and vary in level. Very similar is the script of most sufferers from *writer's cramp.* In *multiple sclerosis* the letters do not run into one another, but the script is shaky, and there are very regularly found deviations corresponding to the tremor in the stroke, the general direction of which is correctly maintained. Of the genuine forms of *tremor, paralysis agitans* has the most typical change in script, the so-called flower script: the easily legible letters in all their parts show, instead of straight lines, small zig-zag lines, consisting of deviations of the length of a millimeter. For the writing of *sufferers from progressive paralysis,* etc., cf. the special chapters concerned.

Pathological *hypertrophies* are much rarer; only acromegaly and pseudohypertrophy in dystrophy are really concerned here. Both likewise give typical pictures.

A symptom of great importance or rather the nucleus of several large groups of symptoms is *muscular tonus.* By this one designates the condition of a certain permanent irritability, so to speak, in the sound muscle (also in condition of rest), which we may interpret as a reflex process localized in the ganglion cells of the anterior horn of the spinal cord. In this process, the anterior horns represent to a certain extent an electrical power station (*Schuster*), which is influenced by the higher centers of the cerebrum as well as by the periphery. Peripheral sensory stimulations may increase temporarily the tonus of the muscles (precisely by way of this power station), as well as may the impulse of the will emanating from the cerebrum, evoking the voluntary movement; on the other hand, there doubtless exist also between the cerebrum and anterior horns inhibiting influences, since in interruption of this conductivity the tonus may increase.

As to the tonus of the musculature, we can get some data from simply testing the musculature, partly also from mere observation of the patient. If his limbs lie flaccid, touching throughout their entire extent whatever they are resting upon, possibly exhibiting also overextended joints, there is present in general, a decrease in the tonus of the muscles; if they lie rigid in more or less forced positions, and no joint fixations can be determined, it is

usually a matter of increase in tonus. The former condition is known as *hypotonia*, the latter as *hypertonia*, of the musculature. Still more obvious is the difference in testing the voluntary, active, and especially in *testing the passive movements*. One instructs the patient to relax his limbs as much as possible and then places the different parts of the extremities now with quicker, now with slower, now with brusque, now with gentle, passive motions into all physiologically possible positions. Nearly always one feels resistance at first, which must be interpreted in part as reflex increase in the normal tonus, and in part is actively, though unconsciously evoked by the patient. But, as a rule, this resistance soon ceases in the healthy individual. Now sometimes, precisely in cases of hypertonia, this decrease does not take place; nay rather, at every attempt at a *brusque* motion, the antagonists, that is, those muscles of the patient, the activity of which is to be suppressed for the execution of the intended passive motion, easily become very tense and afford an almost insuperable resistance to it. On the other hand, the resistance may frequently be overcome by slower, more careful motions, but now frequently a second pathological condition appears: an involuntary spasmodic working of the antagonists (in the *sense* of the voluntary motion), which now suddenly, apparently spontaneously, sluggishly indeed, but extensively, sets in motion the extremity which before lay in immobile rigidity. These symptoms of hypertonia belong to the large group of *spastic* conditions, to which we have to reckon also the *increase of the tendon reflexes*, of which we shall speak in our discussion of reflexes. In spite of the often, as has been said, almost invincible, tension of the musculature in most spastic conditions, its *active strength* is *lowered;* the so-called *spastic paresis* occurs. As a consequence of lasting spastic paresis, we find permanent tonic conditions of the musculature, *contractures*, nearly always accompanied by stiffening in the

FIG. 6.—Spastic paralysis (Little's disease).

regional joints. These contractures of the tense, active (though occasionally also actively paretic) musculature are theoretically (this is more difficult in practice) to be distinguished from paralytic contracture, in which it is a question of mere preponderance of the strength of the antagonists as opposed to the paralyzed agonists that have lost their functional ability (Fig. 6).

Decrease of muscular tonus, hypotonia, is found especially where the stimulations from the periphery to the "power station" (cf. above) of the anterior horn are more or less completely lost, that is in lesion of the sensory tracts above all. Here passive motions may be performed with abnormal ease; the limbs as a result of the relaxation of the muscles (usually also of the articular ligaments) may be brought into the most exaggerated, distorted positions. The knee joints are overextended in walking (genu curvatum), etc. We see the most severe forms of hypotonia in tabes dorsalis. At the same time, the motor strength may be altogether normal. The tendon reflexes are often simultaneously (but by no means always) weakened or absent. Here belongs the remarkable affection, first described by *Oppenheim*, later also by *Bernhardt* and others under the name of *myatonia:* in early infancy, there appears an abnormal flaccidity of the entire musculature, a high degree of hypotonia, while at the same time, direct and indirect faradic excitability is absent. The condition probably represents a retarded development of the central nervous system and can be cured.

If we return again to the test for active motility, we see frequently in nervous patients a disturbance which is not evinced in a decrease of strength or volume, but in the incorrectness of the voluntary movement: the movement becomes inco-ordinated; we have a *disturbance of co-ordination* before us.

The concept of the co-ordination disturbance is to be defined as follows: In every movement of our body (with very few exceptions) several muscles, not *one alone*, are synchronously set in action. The performance of a direct movement is, therefore, an extraordinarily delicate co-operation of these muscles, known as synergists; if only one muscle misses the cue, so to say through too slight or excessive contraction, the entire movement is disturbed. Still more complicated is the process, if (with the majority of authors) one believes that the antagonists co-operate in every voluntary movement of the agonists, whether it be through a particularly delicate gradation of their relaxation or in some other way. If now for some reason the function of one muscle is lost, the others naturally preponderate; the part of the body concerned is then—possibly only for the duration of a fractional part of the entire movement—moved in a direction other than the one desired, until a new impulse sent to a muscle, serving as a corrective, pulls it back again into the old direction. Thus there arises a zig-zag movement, a deviation or throwing about of the extremity concerned; we describe this movement as inco-ordinated or *ataxic*. This disturbance is more or less independent of the will of the patient, but may be somewhat set aside through practice,

and often occurs but slightly in a patient who controls with his eye all his movements, and at every deviation "at once" calls into play corrective muscles, but always increases, if one tells the patient to close his eyes, and thereby removes the "eye-control."

The **test of ataxia** is therefore made as follows in the individual case. First the patient in a position of supination is asked to make complicated movements with his hands: put his forefinger to his nose; thread a needle; perform the so-called "*finger-tip*" *experiment* (the arms which have been drawn back far laterally, are brought together slowly with extended forefingers until they touch). When doing these acts there appear very distinct deviating movements, which produce quite irregular varying twitching excursions, in various planes about the axis of the movement desired (in contrast to the regulated movements of most kinds of tremor), but the patient, with the assistance of the controlling eye, is usually still able to reach the point aimed at. If no clear ataxia has been shown in this, one has the patient perform the same movement after closing his eyes, whereby the ataxia usually increases considerably. To test the legs in ataxia one makes use of the so-called "*knee-heel experiment*": the patient lying down is asked to touch with the heel of one foot the knee of the other leg, by moving the leg through the air (not by drawing it up along the recumbent leg). Then one should have him form letters, figures (three) or draw circles in the air with the extended leg. All these movements should be repeated with closed eyes. An ataxia of the facial muscles, too, may occasionally be established by corresponding tests. Then the patient is to sit up. Here there frequently appears in a quiet sitting posture swaying of the upper part of the body (which still increases upon standing), comparable possibly to a tree shaken by the wind, which also increases when the eyes are closed (*static* ataxia). Now one should have the patient rise and place his heels and toes close together. Very many ataxics fail even in this movement, they begin to totter and have to put one foot on the side, so as not to fall; the disturbance becomes clearer if one has the patient stand on one foot—a thing that a normal person can do easily after some slight preliminary swaying—or specially, if, when standing with his feet close together, he is asked to close his eyes. This uncommonly characteristic symptom (*Romberg's* phenomenon) is one of the most delicate and most easily performed tests of co-ordination, and the manner of the response should be remarked, as it is somewhat different in the various forms of ataxia.

In the *ataxia of tabetics* (the most common variety), the patient first attempts to struggle against the swaying by means of slight movements with his feet; then he places his feet somewhat apart, and finally opens his eyes. In *cerebellar ataxia* (cf. below) the swaying, already in itself of high degree, does not increase when the eyes are closed. In the ataxia of hysterics (especially in *traumatic hysteria*) the swaying becomes noticeable only when

the eyes are closed, but then immediately becomes excessive; the patients make no attempt to arrest the interesting disturbance and fall with apparent carelessness (but in reality with great caution) to the ground.

In *walking*, ataxia, as a rule, does not become higher in degree, than in standing, but it assumes peculiar forms. The ataxic tabetic walks (in the most severe cases) with legs far apart, protects himself, so to speak, by planting his feet as widely apart as possible (to overcome the swaying that seizes him) like a sailor on ship-board. He likes to use a cane, which he puts down as far as possible from him, in order to gain as large a triangle as possible to serve as a firm support. Other tabetics maintain the direction of their movements, but raise their legs (usually in extension) unnecessarily high, and replace them with stamping, a kind of walk, recalling the older forms of the German military goose-step, usually called "swinging" gait. The "cerebellar ataxic" walks quite differently. He totters like a drunken man; the working of the synergisms necessary for walking is normal, but the movements are wrongly connected with one another; the gait looks as if the patient had been suddenly seized with vertigo (which, as a matter of fact, is often connected with this form of ataxia). To the group of cerebellar ataxia belongs also the disturbance described by *Babinski* as "*asynergia*," which occasionally appears unilaterally. Ataxic tabetics walk, as a rule, far more poorly in the dark than by day; this statement also is often characteristic, that their ataxia had been noticed first by them in the morning when they had closed their eyes to wash the face.

The regulation essential for co-ordination of movements is mediated by centripetal—in the main, therefore—sensory stimulations, which become transformed in the centers of the cerebrum and the cerebellum; it is not yet known how far the system of semicircular canals in the labyrinth, which unquestionably serves to maintain equilibrium, subserves *co-ordination.* Disturbances in co-ordination, ataxia, will appear, therefore, as soon as the paths or centers of regulation are interrupted: tabes, *Friedreich's* disease and possibly ataxic polyneuritis on the one hand, affections of the cerebellum and (very rarely) those of the cerebrum, on the other, lead to ataxia, but, strange to say, never mere affections of the centrifugal path—that is, pure motor paralyses with exception of the rare polyneuritic ataxia. We must, moreover, here repeat again that ataxia has nothing to do with disturbances in coarse strength, though occasionally, motor conditions of weakness other than ataxia may occur (ataxic polyneuritis).

Ataxia, to a certain extent, represents an "insufficiency" of irritation phenomena of the motor sphere, combined with *unregulated* motor innervation. We come now to several symptomatic groups, all of which are characterized by an "excess" of motor irritation phenomena.

As *tremor* are designated regular involuntary movements following

one another rapidly, taking place in smaller or larger excursions about an axis and always in *one* plane; they are evoked by involuntary muscular contractions, in which agonists and antagonists alternately participate. One distinguishes between a fine and a coarse tremor, according to the size of the excursions, eventually also between a rapidly and a slowly oscillating tremor. Furthermore there must be differentiated, principally, a tremor which appears only upon motion, and one that appears only in rest. The two extremes are represented by the tremor of multiple sclerosis and that of paralysis agitans.

The sufferer from multiple sclerosis in a state of rest, is, as a rule, absolutely free from tremor. Also in simple tonic innervation, if, for instance, he holds his hands quietly extended, no tremor appears. As soon, however, as he intends to perform complicated movements, as in the test in which an

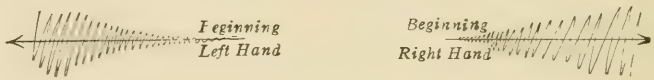


FIG. 7.—Tracing of the intention tremor in multiple sclerosis.

attempt is made to bring together the tips of his fingers, tremor appears. This tremor is first characterized by small excursions, but the more the movement comes "to a point," the more delicate it becomes, the greater the demands to reach the desired point, just as in the finger-tip experiment, at the moment the finger tips meet, the larger the excursions become, which can be represented by a diagram as follows (Fig. 7):

If one has an ataxic perform the same movement, the diagram will be something like the following (Fig. 8)—in which one must consider, moreover, that the deviating excursions of ataxia may lie in altogether different planes.

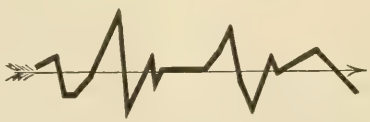


FIG. 8.—Tracing of intention ataxia in tabes. The individual strokes of the line frequently lie in quite different planes.

It is, therefore, certainly not permissible to confound the "*intention tremor*" of multiple sclerosis, with the "*intention ataxia*" say, of tabetics, though there may be some points of similarity. Even the innervation disturbances upon which they depend may be different.

Quite different is the tremor of *paralysis, agitans* (*Parkinson's* disease). Here in intended, voluntary movements, there is little or no tremor; but if the patient allows his extremities to lie or hang quietly, if he places his hands in his lap, etc., there appears usually a very fine tremor with rapid oscillations, which, in severe cases, persists in sleep and in the beginning often attacks only one extremity, then, perhaps, half of the body, finally,

usually the entire body. It also appears in the hands in a peculiar type of tremor, designated as "coin-counting" (pill-rolling), which consists of movements of the thumb in front of the other fingers—that recall the movement from which it derives its name.

Most other forms of tremor are of the type of fine oscillations, usually only slight but nevertheless present in the state of rest, increasing upon motion, such as *neurasthenic* and *hysteric* tremor, *alcoholic* and *senile* tremor and the tremor in *Basedow's* disease. Slight tremor can be made plainly visible by laying a piece of paper on the outstretched fingers of the patient.

Nystagmus represents, as has been mentioned above, an "intention tremor" of the eye muscles. Some forms of tonic spasm, as, for instance, blepharospasm (eye-lid spasm) of hysterics, are also associated with slight, finely oscillating, tremor of the muscles concerned.

The so-called "*fibrillary*" or "*fascicular*" *twitchings* of parts of one muscle represent a totally different process. In this, we see at various places on the surface of the body, small twitchings of individual bundles of muscles, usually taking place without any effect on movement; they are occasionally rhythmical, recalling, then, especially shivering from cold, but are frequently arythmical, attacking always different muscular bundles in the same region and may lead to a total "muscular wave" (*myokymia*). The genuine fibrillary, that is, smallest, muscular twitchings of this kind are found only in organic diseases of the *central* nervous system. All other muscular groups can, after sufficient practice, be distinguished from these. We must add here, moreover, that in affections of the anterior horn, etc., which are associated with muscular atrophy, these twitchings are restricted, in the main, to the atrophic areas, and occasionally also indicate the spread of the process to other muscular parts preceding the atrophy.

In the realm of involuntary movements, there belong the *associated* or *co-movements*. Small children scarcely ever make movements of grasping, etc., with one hand alone, but innervate both hands and arms synchronously; complicated processes, such as crying and laughing, are also very frequently found together with associated movements on the part of the extremities. These co-movements, with the progress of development disappear, leaving behind only traces, but are occasionally found even at a more advanced age, and return in pronounced fashion in organic paralyses, especially in those of cerebral origin. Most hemiplegics, for instance, can not make energetic movements of the sound arm entirely without associated movements of the paralyzed arm, and this symptom is an important differential diagnostic criterion between organic and psychogenous, hysteric paralyses, in which latter it is always lacking (*Hans Curschmann*).

Finally the last three groups of involuntary hyperkinesis, those which we designate choreic disturbance in movement, tic and athetosis, show a considerable similarity to one another. Each is well characterized in itself,

presents a peculiar form of disease, may often be classed among the neuroses, and may therefore occur without any "organic symptoms." Therefore their knowledge and recognition are particularly important. *Chorea* is characterized by sudden violent twitching excursions, without rhythm and regularity, appearing in rest, and not totally disappearing upon motion. In these movements single muscles as well as muscular groups and entire extremities are involved; in the general impression the "restlessness of the limbs" predominates, since some of the violent movements may seem absolutely intentional. The patient puts his hand up to his nose, purses his lips for whistling, reaches for an object, etc. Frequently the disease is first unilateral, but usually it appears over the entire body affecting especially, in most cases, the face (making grimaces, rolling the eyes, etc.)—and besides this, very frequently the psyche also is affected (Fig. 9). In the most severe cases, the twitchings follow one another in uninterrupted succession, the patient throws himself about in bed, tears his hair, rends the sheets and his clothes, can scarcely be quieted with hypnotics, and frequently dies in this condition. Less severe are the comparatively rare choreic movements in organic diseases of the brain. In contrast to this, *tic* represents a regulated movement, in so far that it always attacks only definite groups of muscles, usually, however, in the same brief twitching way as in chorea. The movement may, at first sight, appear voluntary, intentional; the patient shrugs his shoulders, throws his head backwards, strikes the table with his hand. The same movement is repeated, not rhythmically, but at all times, upon all occasions, increases especially during excitement, and can not be suppressed in spite of the most earnest desire of the patient to put an end to it. If the motility disturbance affects muscular groups over the entire body, one speaks of "maladie des tics," which, however, can not, in its nature, be totally identified with simple tic (*Gilles de la Tourette*). Chorea and tic are frequently accompanied by psychic disturbances. *Athetosis* also occasionally occurs as

FIG. 9.—Clonic spasms in chorea hereditaria. (*After Huntington.*)

an independent neurosis, sometimes, however, after hemiplegia in the paralyzed limbs, is frequently combined with contractures and nearly always affects only fingers or toes. In this case, the movements are sluggish, vermicular, always with considerable effect upon motility and often of very strange form. The fingers are extended, stretched, overstretched and bent in the most remarkable manner, assuming the most unlikely positions. Whereas in chorea and especially in tic there are usually longer or shorter intervals of rest between the separate movements, athetosis is an almost unceasing hyperkinesis, suppressed, as a rule, only during sleep.

Spasms (cramps) have often been designated as hyperkinesis in the narrower sense, among which we can with some justification reckon the motility disturbances we have just mentioned (especially tic). We distinguish as main groups, *tonic* and *clonic* spasms. By tonic spasm we mean permanent conditions of contraction in one muscle group; the most severe forms are called tetanic spasm, tetany. Clonic spasms are brief, rapid twitchings, usually repeating themselves with a certain rhythm, which may, in the highest degree, attack the entire body (convulsions). We have already found an example of isolated clonic spasm in tic. It appears frequently as a reflex spasm (tic convulsif of the facial area). The majority of spasm forms belonging to this chapter, however, are to be classed among the neuroses or psycho-neuroses, and may all probably be localized in the cerebral cortex. This is certainly true of the spasms in *Jacksonian* epilepsy, where, as the result of irritation of a definite part of the cortex (scar, tumor) tonic, or more frequently, clonic spasms, always of definite muscular group, usually only unilateral in the opposite half of the body, are evoked, with or without loss of consciousness. Analogously, we may assume that convulsions of common *epilepsy* may originate in the cerebral cortex. The question of localization with respect to the major *hysteric* attacks of spasms is more difficult to answer (Fig. 10). In appearance, epileptic and hysteric attacks are but little different and may pass over into each other. As to the differential diagnosis, more will be said in the special chapters. In general, the tonic conditions predominate in epilepsy; usually consciousness is entirely lost. In hysteria, even the "major attack" is usually composed of periods of tonic rigidity and clonic twitchings and consciousness is not totally lost, at least a psychic reaction can usually be evoked by means of external influence, especially if painful. The *epileptic* patient falls to the ground, usually without noticeable prodromes (aura) of the attack, his arms and feet twitch a little, his teeth gnash spasmodically, and there soon appears a high degree of tonic tension throughout his entire body with finely oscillating clonic vibrations of the muscles in tension; respiration becomes superficial, the patient wheezes, becomes cyanotic, foams at the mouth, frequently urine is voided involuntarily, until after a few minutes (rarely more), tension is relaxed and consciousness as a rule, soon returns with the restoration of the reflexes that had dis-

appeared (also of the pupils). The entire event creates an impression of a real, genuine attack; not rarely the patients are injured by falling, or bite their tongues. In the *hysteric attack* half is usually "histrionic." The attack rarely appears when the patients are wholly unobserved; frequently it crowns the demonstration of a passionate emotional outbreak. The hysterical patient produces his effects with far coarser means than the epileptic; the entire body is convulsed with twitchings, assumes "attitudes passionelles," like the position of a crucified man, erotic positions, etc., lies in bed in the form of an arch, so that only the back of the head and the heels touch the

FIG. 10.—Hysteria (major hysteric attack). Attention is called to the opened and fixed eyes. (*After Schoenborn-Krieger, Klinischer Atlas der Nervenkrankheiten.*)

sheet. Any suggestive influence, pressure in "hysterogenous" zones, faradization, in lighter cases, sprinkling with cold water and addressing the patient may check the attack, which otherwise may last for hours. If the patient falls to the ground, he usually falls carefully so as not to injure himself. In spite of all these signs of recognition, the differentiation of these two forms of spasm may, as has been said, be very difficult. For a discussion of separate rarer forms of spasms, such as the tonic *tetanic* spasms which are localized especially in the extremities, *myoclonus*, occupational cramps, as well as for traumatic *tetanus*, cf. the particular divisions devoted to these subjects.

In the test of motility or of coarse strength we have still two phenomena to mention. In some cases, upon first testing the muscles, as in warmly shaking the patient's hand to see how strong his grip is, etc., his strength is

very good, but he is unable to relax the muscle that has been contracted. But if he repeats the same movement several times in succession, "for practice," the contraction and relaxation take place more and more rapidly, finally, at the normal rate of speed; the grasp of the hand, which, like the clutch of a vise, would not, in spite of all the endeavors on the part of the patient, release the examiner's hand, can now no longer be distinguished from the grasp of a healthy man, until after some period of rest, the first condition reappears. In this disease, *myotonia* (*Thomsen's disease*), which, as a rule, attacks the entire body and may be accompanied by very good coarse strength, even with hypertrophic musculature (cases of atrophy are rare), abnormal anatomic conditions and transformations in the muscle itself probably play the main ætiological rôle. In a certain sense, *myasthenia or myasthenic paralysis* forms a contrast to this—the picture of an abnormal tendency to fatigue of the otherwise apparently normal musculature. Here, it is true, the patient at first, for a few times, may perform the prescribed movement rapidly and powerfully; after the fifth or tenth time his strength decreases, and soon, in spite of all his efforts, he is unable to execute the movement, until, after a resting period, the muscles have again recovered. For this disease picture, which has not yet been consistently interpreted, there exist, moreover, as for myotonia, analogous electrical changes (cf. below).

An absolutely essential part of every test of the motor sphere is the *electrical examination*. But it should be preceded by the test of the *mechanical excitability of the muscles and nerves*, comparable with it in many respects. We understand by this, naturally, not the pain stimulations that affect the skin, but the movements due to pressure, tapping or striking, which affect the skin as little as possible, and strike the underlying portions, as far as possible, in isolation. For this test, almost without exception, only the finger of the examiner or the tap of the percussion hammer is suitable.

Changes in mechanical muscular excitability are demonstrable almost exclusively in the form of an increase in excitability, or of qualitative change in it. For there is no norm in respect to the effect of tapping a sound muscle. The local muscular prominence may form an "*idiomuscular contraction*," but it may be very slight and, as happens in the majority of cases, may disappear again immediately; it may also be very pronounced and persist for some time. This *increase in mechanical muscular excitability* is found in tuberculosis, typhoid, in most cachexias, here and there, in sciatica and affections of the anterior horn. Very frequent, and occasionally classical, on the other hand, is the qualitative change in excitability, the mechanical proof of the reaction of degeneration (cf. below), or more correctly the "sluggish mechanical contraction" as it is often found in the degenerative atrophies, especially at an early stage. In ulnaris neuritis, bulbar paralysis, etc., tapping the ball of the little finger, or the thumb, evoke the most beautiful

"sluggish contractions." In Thomsen's disease, also, the mechanical tonic twitching corresponding to the myotonic reaction (cf. below), the "arrest" of contraction may be typically demonstrated. The theoretically postulated *lowering* of mechanical excitability for severe old paralyses, for myasthenia, etc., can not, on the other hand, as a rule, be shown. For the *mechanical excitability of the nerves* also—which, in our sense, can be used only for motor nerves—only *an increase in excitability* can be demonstrated; decrease and qualitative change can not be proved. The mechanical increase in the excitability of the nerves is best shown in some nerves lying near the surface:

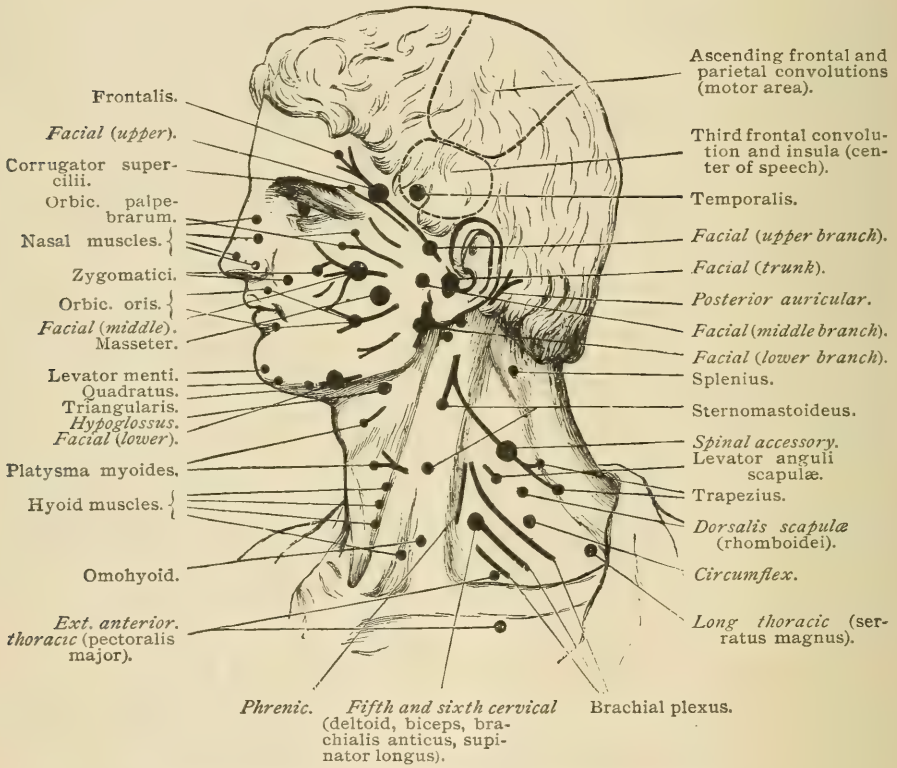


FIG. 11.—Motor points of face and neck. (*After Erb and de Watterville.*)

ulnaris (at the elbow), peroneus (capit. fibulæ), facialis (most of its branches as well as the trunk). This increase, as a matter of fact, is found only in a single disease, tetany, in which tapping upon the ulnaris at once evokes twitchings in the muscles of the ball of the little finger, tapping or stroking of the facial branches, unilateral twitching of the mimetic muscles (*Chvostek's symptom*).

The conclusions reached by testing the *electrical reaction of the muscles and nerves* are very much more comprehensive. But in this field, which in modern medicine, even in neurology, is somewhat unkindly treated and

rather neglected, there are necessary, more than in others, careful observation, experience and detailed knowledge of the technique (above all of the electrical apparatus used by the examiner). Therefore, if possible, one should make an electrical examination of patients, only with an apparatus one owns

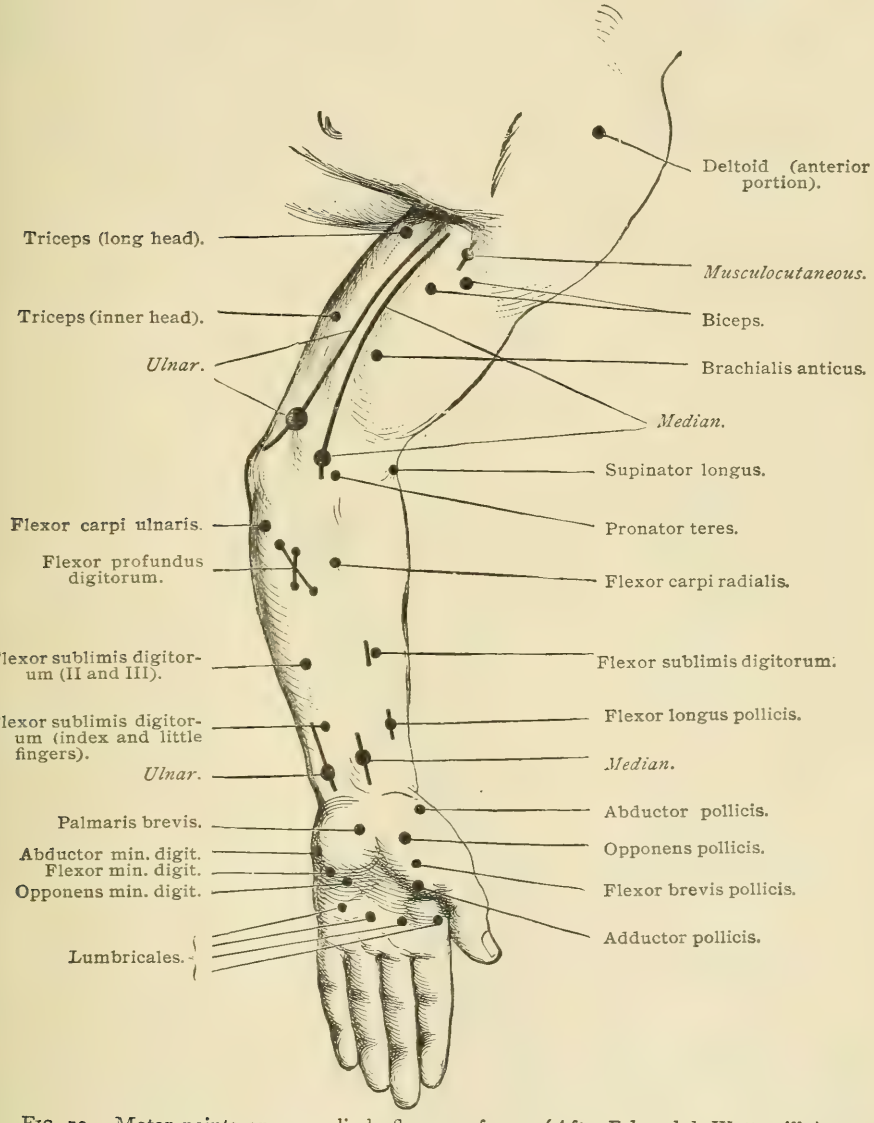


FIG. 12.—Motor points on upper limb, flexor surface. (*After Erb and de Watterville.*)

oneself, or, at least, always with the same kind, which should comprise a faradic (with the greatest possible variation in the relation between tube and core) and a galvanic battery with a good galvanometer, as well as several ordinary electrodes, a button electrode and several plate electrodes (surface of 10 to 60 sq. cm.).

We work, as a rule, with two electrodes of different size, a large so-called indifferent one, about 60 sq. cm. and a small active one, as like as possible Erb's normal electrode of 10 sq. cm. (round or square). The large one is placed upon an indifferent spot (the sternum is best) and may be held there by the patient himself; the small one is placed upon the muscles and nerves to be tested. For the necessary opening and closing of the current, one makes

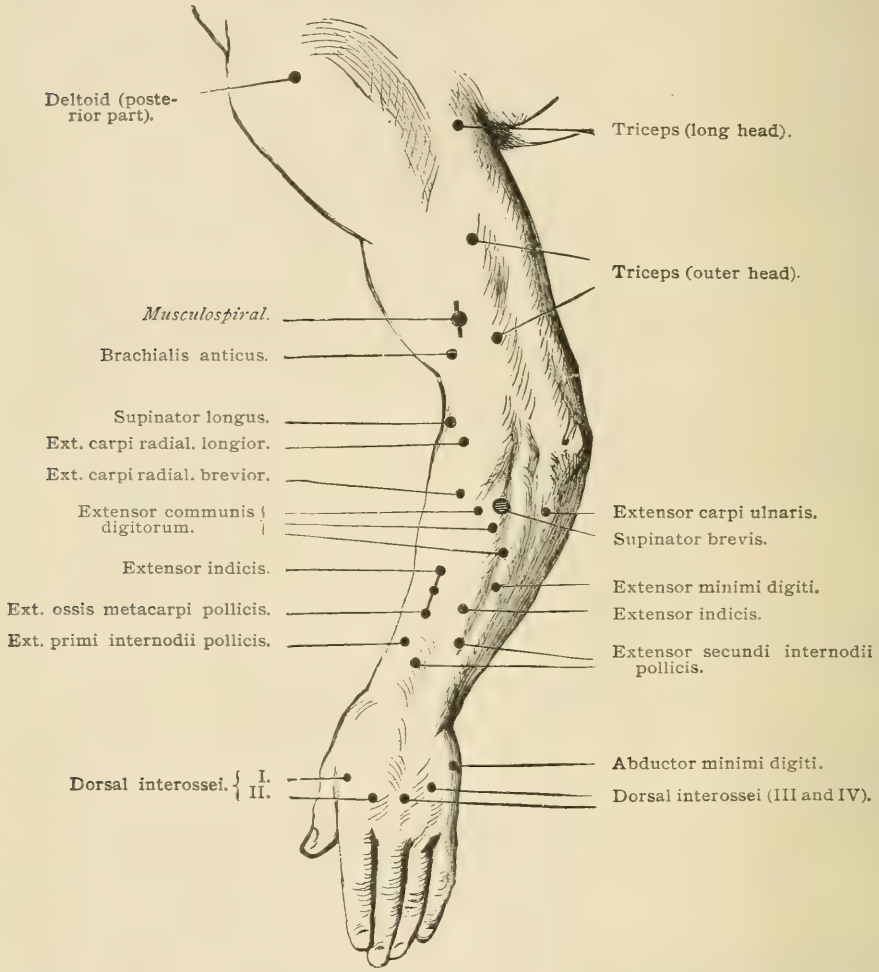


FIG. 13.—Motor points on upper limb, extensor surface. (*After Erb and de Watterville.*)

use of the interrupting electrode, upon the handle of which, pressure on a button or a smiliar device, causes a break in the current.

Now, *which points* does one examine? Where does one set the active electrode? In general, upon the empirically determined points of entry of the nerves into the muscles—for the electric test of the *musculature*—and upon the places in the *nerve* trunks where these approach the surface. These "muscle and nerve points" were determined by *Erb* upon a foundation of

extensive and detailed investigation (Figs. 11 to 16). The significance of the individual points is clear, without further discussion; they show that if one places the large electrode upon the sternum and the small one upon one of the points upon the surface of the body here designated and employs (faradically) a sufficiently strong current or (galvanically) opens and closes the current, then, the muscle named in the tables (at the nerve points, the muscles subserved by this nerve) twitches, that is, momentarily (galvanically) or tonically, lastingly (faradically) contracts. If it be a matter of a diffuse affection, all these muscular and nerve points must be examined in this way,

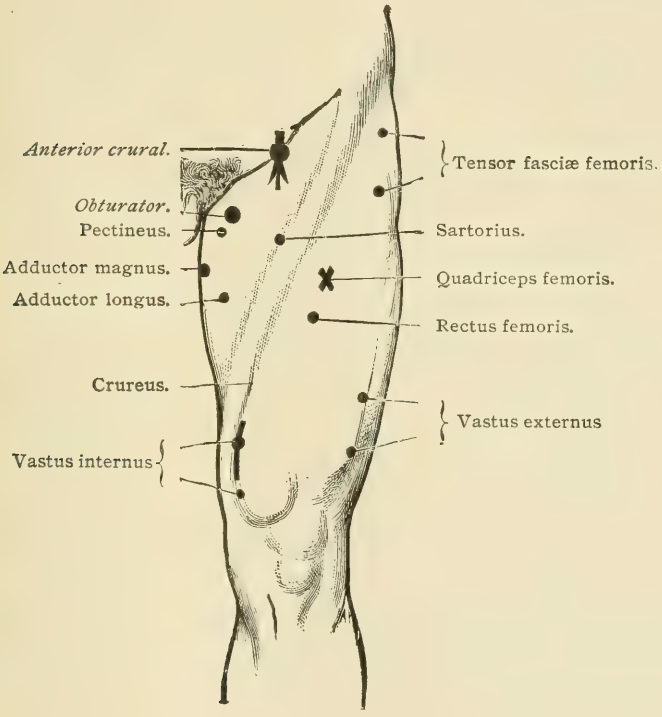


FIG. 14.—Motor points on thigh, anterior surface. (*After Erb and de Watterville.*)

while in local affections only the regionary muscular and nerve points are to be thus examined. First the faradic and then the galvanic examination should be made.

In testing the faradic excitability, one determines what minimal strength of current barely suffices to evoke a contraction of the muscles, and (theoretically) in the *indirect* test—stimulation of the nerves—as well as in the *direct* stimulation of the muscles. This minimal power of the current is shown in millimeters upon the instrument, but since all apparatus for measuring the faradic current are unreliable, and not absolutely equal in any two sets of

apparatus, the examination is of direct value only where one can compare in a patient the diseased muscle and nerve with the sound one on the other side, when one can, to a certain extent, estimate the capacity of the skin to resist the current and works with an apparatus one has already tried. Certain normal medium figures for these "threshold" values have been determined by *Erb*, *Stintzing* and others and may serve as points of reference; usually *Stintzing's* figures are used. But the unskilled examiner (especially when the nerve points are, as is sometimes the case, hard to find) often meets considerable variations from these mean values. Of these mean values, we shall reproduce here some from *Stintzing's* instrument. But we must emphasize the fact, that in direct muscular stimulation, the excitability fluctuates within such wide bounds, that usually one is contented with reproducing the most manageable values of the limit of indirect (nerve-) excitability.

Nerve or muscle	Distance table for minimal twitching
N. facialis	132–110
N. medianus	135–110
N. ulnaris	130–107
N. peroneus	127–103
N. cruralis	120–103
N. radialis	120–90
N. accessorius	145–130
N. musculocutaneus	145–125
N. axillaris	125–93
N. thoracicus ant	145–110

The direction of the current is of no importance in this test. In faradic stimulation, upon interruption of the current ("opening shocks") there appears a momentary contraction, otherwise, a tonic contraction lasting as long as the current is applied.

The procedure in *testing the galvanic excitability of the muscles and nerves* is somewhat different. Here, too, the large indifferent electrode is placed upon the sternum (possibly upon the nape, or the lumbar portion of the spinal column), the small, active one upon the points to be tested. Only opening and closing of the current evokes muscular twitching, *not* the duration of the current. The contraction in the normal state, is brief, lightning-like; tetanus appears only upon the use of very strong currents—but never "sluggish contraction" (cf. below). But here the direction of the current is of importance. We always begin by taking as the "*active*" electrode, the *cathode* or *negative* pole.

Now we determine at what power of the current appears the minimal

contraction for direct and for indirect stimulation. The measure of the strength of the current is here given exclusively by the *galvanometer*, which directly indicates in milliamperes the strength of the current. It is best, in every examination to insert the galvanometer from the first into the circuit of the current, since the resistance that it always offers to the current, would, if it were put in and taken out later, destroy the result of the test; if the galvanometer is excluded, a smaller number of elements will always be necessary to secure the same strength of current and the minimal con-

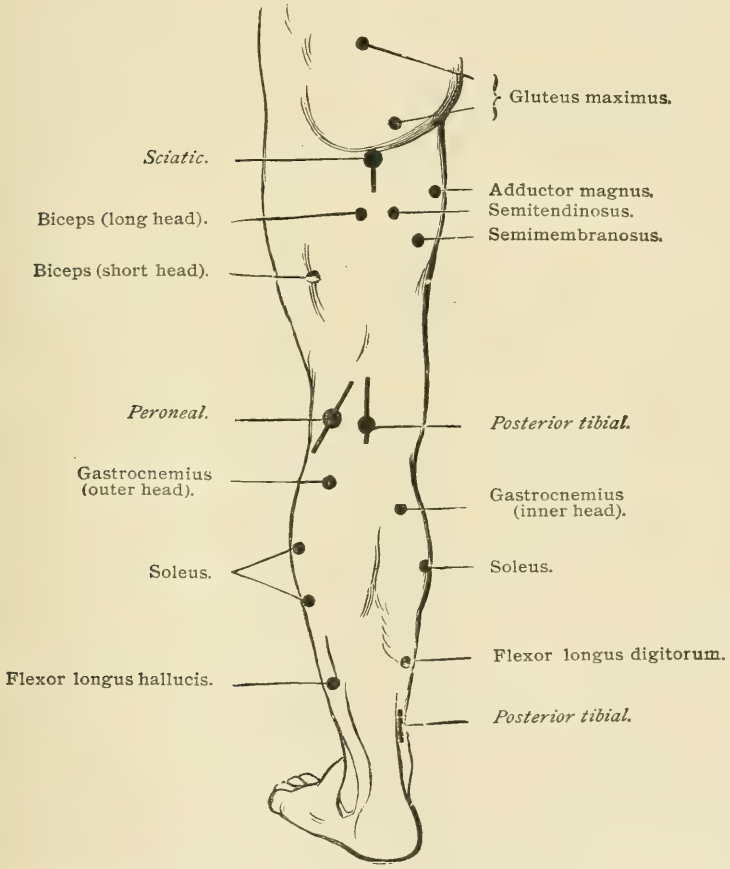


FIG. 15.—Motor points on lower limb, posterior surface. (*After Erb and de Watterville.*)

traction. Furthermore, one must see to it that the electrodes, as always should be done in electrical examinations, are thoroughly moistened with warm water (salt water is unnecessary), and remember that in the galvanic test, the resistance of the skin to the current decreases with the length of application.

For galvanic minimal contractions, too, a series of average values have been given; some of the most important, according to Stintzing, are:

Nerve	Minimal strength of the current in Milliamperes (mean wave)
N. facialis	1.0 –2.5
N. accessorius	0.01–0.44
N. medianus	0.3 –1.5
N. ulnaris	0.6 –2.6
N. radialis	0.9 –2.7
N. cruralis	0.4 –1.7
N. peroneus	0.2 –2.0
N. tibialis post	0.4 –2.5

The values, as shown above, vary considerably even in sound persons. But the following must be remarked: these values, and especially the

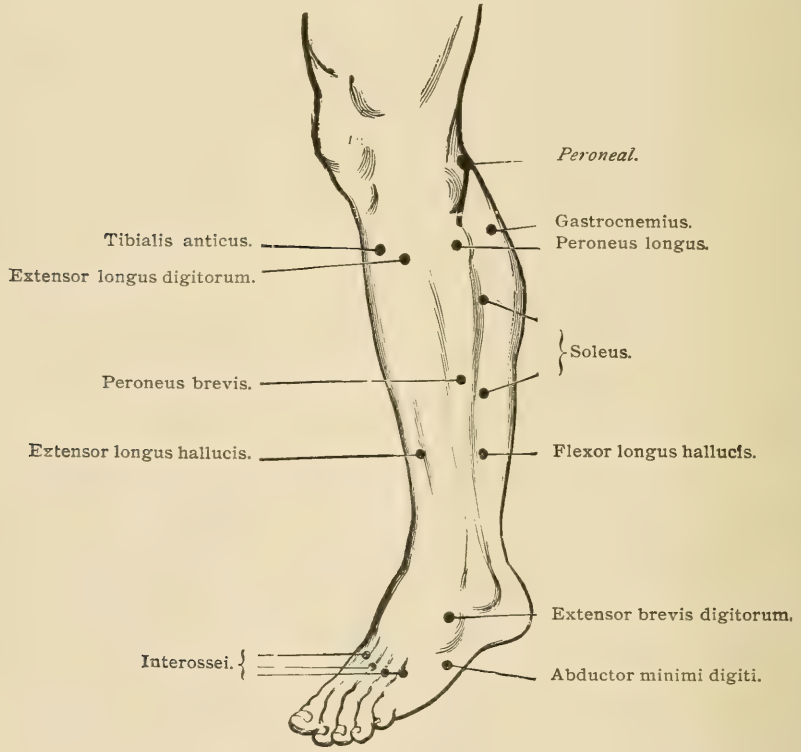


FIG. 16.—Motor points on leg, external surface. (*After Erb and de Watterville.*)

"threshold" values of the galvanic stimulation of the sound nerves, hold good for the *cathodal closure contraction* (C C C) that is, for the twitching, which appears when the cathode is placed as active electrode upon the point to be tested, and the current is passed in, then closed off. According to the *law of contraction* this is the twitching that first appears in normal nerves.

Now, if we open the current, and then by means of the commutator which is necessary in every galvanic battery, reverse it, and then alternately interrupt it (opening and closing), whereby the active pole which was formerly the cathode has now become the anode, we find, if the power of the current is kept as strong as before, no further twitching. But if we slowly increase the current, we see first that, besides the twitching that results from cathodal closure and remains the strongest, there appears a contraction upon opening (interrupting) the current, at the pole that has become the anode—the *anodal opening contraction;* at the same time, too (sometimes a little earlier), there occurs a *contraction at closing the anode* (A O C and A C C). If we further increase the current, there results at the *cathode,* upon closing the current, instead of the former brief twitching, *tetanus* (C C Te), as long as the current is kept closed, upon still further increase, *contraction at the opening of the cathode* (C O C), and an *anodal-closure tetany* (A C Te). Naturally, it is theoretically possible and occurs also in practice, that at the place where the indifferent electrode is used, twitchings are also evoked simultaneously with those at the active pole (occasionally especially disturbing in facialis paralysis). This disturbance, which may make the picture confusing, should be avoided by taking a large electrode at the place which one wishes to be indifferent (at which if possible no contractions are to be made manifest), which possesses slighter *density of current,* and by placing this electrode precisely upon indifferent places (sternum, spinal column), where no nerves are located directly below the surface of the skin. This "loop of current" from the indifferent electrode may really become disturbing—if it be a question of determining precisely the place of lesion of a nerve by electricity, and one follows the trunk of the nerve with the indifferent electrode while the active one is placed upon the muscle, so that under certain circumstances the two electrodes approach each other closely.

It should be briefly mentioned that an electrical (as a rule only galvanic) test of the *sense nerves* may be performed. The examination of the *eye* is made as follows: the indifferent large electrode is placed upon the sternum or nape, the active, small (normal) electrode, best covered with a sponge, is placed on the closed eyelid, and now the current closed, naturally with weak current values. The law of contracture remains as above, the reaction of the nerves is an intensive light (sometimes also color) sensation; the threshold of stimulation is usually very low. For the *ear*, it is best to use as electrode, a normal or a button electrode, placing this upon the tragus; there appear buzzing or whistling auditory sensations (the method is much used nowadays for otological tests in labyrinthine vertigo, so-called voltaic vertigo). Finally the sense of *taste* may be tested (small active button electrode on the tongue or cheek), upon which the stronger sour taste appears at the cathode, the weaker salty taste at the anode. This fact,

moreover, may be used to ascertain the position of the pole, if one does not know it; one places an electrode on each cheek, and passes a weak current through; the side upon which a usually distinct sour sensation appears, corresponds to the positive pole, to the anode.

Quantitative changes in electrical excitability of the muscles and the nerves in unhealthy conditions are alone shown when the faradic current is used; nerves and muscles are less or more excitable than in the normal condition; with the galvanic current, on the other hand, quantitative and qualitative changes are shown. We shall now take up the former.

Diminution and even cessation of *faradic* excitability is found for the muscles as well as for the nerves in all possible diseases of the peripheral neurons. Its combination with qualitative changes in galvanic excitability, as in the reaction of degeneration, will be mentioned below. *Mere diminution* is found in all simple (that is not degenerative, cf. below) atrophies, in some cerebral and spinal paralyses, in dystrophy, occasionally too, in other diseases that lead to degenerative atrophy (neuritis, poliomyelitis, etc.), and then, as a rule, as the first or last stage of the electrical change; finally also in the rare myotonia (cf. above). An *increase* in faradic excitability is found almost exclusively in tetany and even there not constantly. Increase or decrease in faradic excitability is recognized simply by the increase or decrease of the gradations for the minimal twitching, as compared with the mean values.

Increase in galvanic excitability is found (except in the beginning of R. D., cf. below) also only in tetany, and here constantly for the excitation of the motor, frequently for that of the sensory and sense nerves (especially of the acusticus); it is recognized by the small power of the current necessary to produce the minimal twitching. *Decrease in galvanic* excitability is found simultaneously with that of faradic excitability in the diseases mentioned above (at least for the indirect stimulation—that of the nerves).

More important, and in certain diseases far more common, is the appearance of the *reaction of degeneration* (R. D.). In this we must distinguish clearly between the electric condition of the nerves and that of the muscles. In the "*total*" reaction of degeneration:

(a) The nerve is excitable neither faradically, nor galvanically.

(b) The muscle is not excitable faradically, on the other hand its galvanic excitability is increased upon direct stimulation (only at the beginning of the disease); besides this the law of contraction is so changed for the muscle that there appears at first reaction (to the weakest stimulation), a twitching upon anodal closing or even upon its opening ("*reversal of the law of contraction*"). Besides this, the twitching becomes *sluggish*, that is, the contraction rises gradually to its height, and then sinks again gradually, slowly; the difference in the twitching curve in direct stimulation of the muscles is about that seen in Fig. 17.

There is also a *partial* reaction of degeneration, generally in less severe lesions, or as preceding or following a stage of total lesions; here the only faradic muscular excitability is decreased or even lost, and sluggish twitching is demonstrable; on the other hand, the excitability of the nerves may be retained for both kinds of currents. The distinction of the total from the partial R. D. may be important diagnostically as well as prognostically, since the latter trouble is cured considerably more quickly.

The reaction of degeneration occurs everywhere, where *degenerative* processes exist in the anterior horns, nerves, or muscles. Here belong poliomyelitis ant., syringomyelia, tumors, myelitis, bulbar paralysis, spinal muscular atrophy, amyotrophic lateral sclerosis; all peripheral nerve lesions; finally in rare cases, some muscular diseases, as to the genesis of which we have not yet full knowledge (dystrophy).

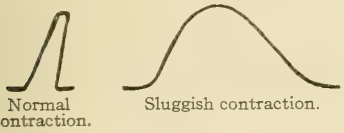


FIG. 17.—Diagram of the normal and the sluggish contraction in closing the galvanic current.

We must mention, that, as excellent investigations of *Grund* have confirmed, the *influence of cold* may evoke the picture of partial reaction of degeneration (sluggish twitching, equalization and even preponderance of the anode over the cathode) ("*cooling off reaction*"). The curves of these twitchings are not identical with those of the R. D., but even exceed them in sluggishness. This "cooling off reaction" may appear spontaneously in the small muscles of the hand of the healthy, and persist for a very long time (for hours even in a very warm room), evince itself also in the larger muscles of the body upon a particular cooling off, and it may appear precisely in diagnostically important cases, in which, then, warming of the muscles is necessary to ascertain whether a partial R. D. is present.

In spite of this possibility of a confusion, the diagnostic value of R. D. is very great; we must emphasize again the special importance of recognizing the sluggish twitching.

The *myotonic reaction* of Thomsen's disease (myotonia congenita) presents a peculiarly abnormal disturbance in electrical excitability. Here, upon direct galvanic and faradic stimulation of the muscles, there appears even with weak currents, a "persistent" tonic contraction of very considerable duration; the My. R. is especially striking upon weak galvanic stimulation. Stronger faradic currents effect also, on the part of the nerves, a tonic, permanent contraction. The phenomenon is demonstrable in the (usually) hypertrophic, as well as (in the rare cases of this type) in the atrophic, muscles of such patients, occasionally very similar to R. D., but always without reversal of the law of contraction.

As "*myasthenic reaction*" (Mya. R.), one designates an abnormal tendency to fatigue, for the faradic current, on the part of the nerves as well as of the muscles, appearing in myasthenia gravis pseudo paralytica. If one

repeats at short intervals, faradic stimulations of equal strength, after some (possibly 8 or 10) stimulations, a rapid decrease, even a disappearance of excitability, will be observed. In rest, the muscle and nerve soon "recover" again. Upon long lasting faradization, the contraction decreases from second to second and finally disappears (*Jolly*).

My. R. and Mya. R. correspond throughout to the analogous changes in active and mechanical excitablity in myotonia or myasthenia. Some rare electric changes will be discussed in the separate chapters.

D. Sensibility

The disturbances in sensibility in their various forms may be subjective and objective, but in no process in the human body is there a greater lack of harmony between the subjective troubles of the patients and the objective

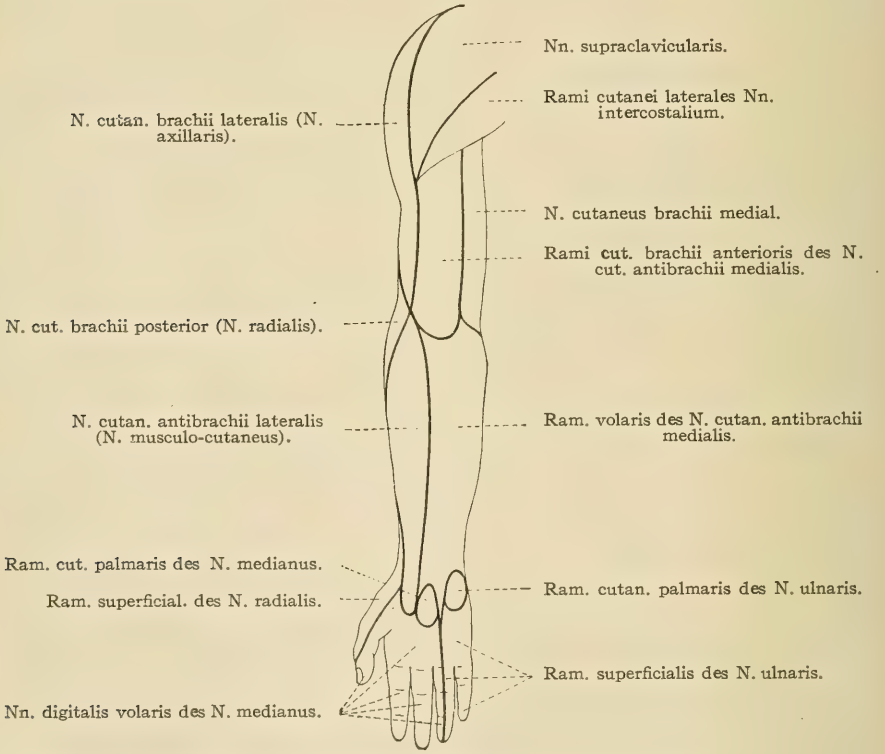


FIG. 18.—Distribution of the cutaneous nerves in the upper extremities. (*After Toldt.*)

findings, than in disturbances in sensibility; nowhere do the deficiencies in our technique appear more sharply than here (Figs. 18 to 24).

A glance at the way *sensibility is tested* will confirm this. We distinguish first, quite roughly, the methods of testing the separate qualities of sensation (the designation "quality," however, has by no means been proved justifiable

in every direction); common to all, is naturally the great defect, that in making the test we are (with a few exceptions) absolutely dependent upon the patient's statement.

We test first the *sense of touch*. We have the patient close his eyes, or cover them, and touch in succession with our finger tip the various portions of the surface of the body. I consider this simplest of all methods the best, because it alone affords the examiner possibility of certain control of the pressure in the experiment, and alone permits keeping touch at the same pressure. In the otherwise much used test by means of a small brush, the

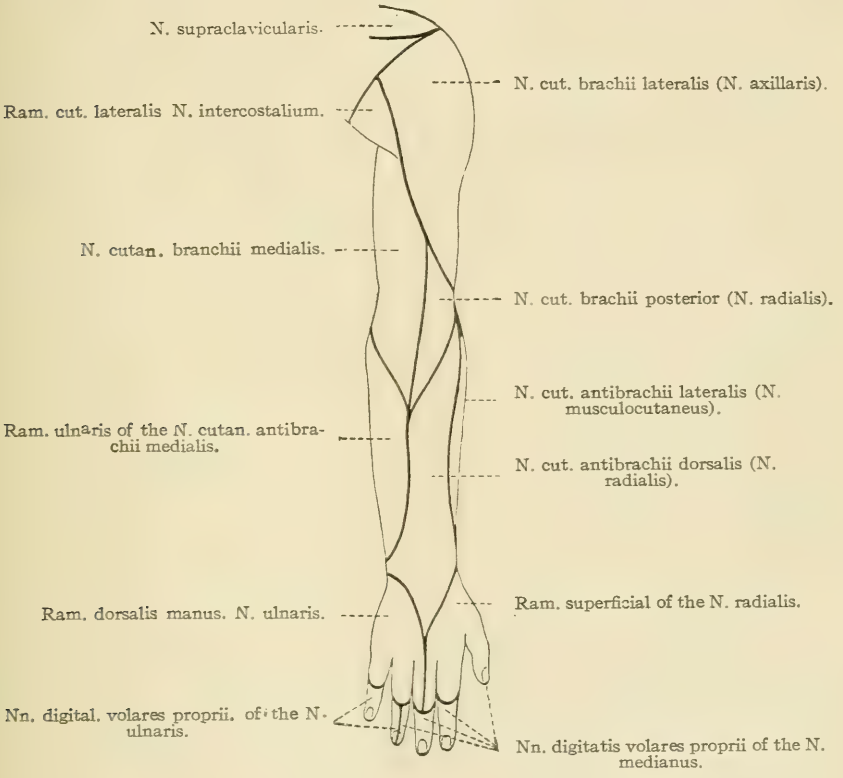
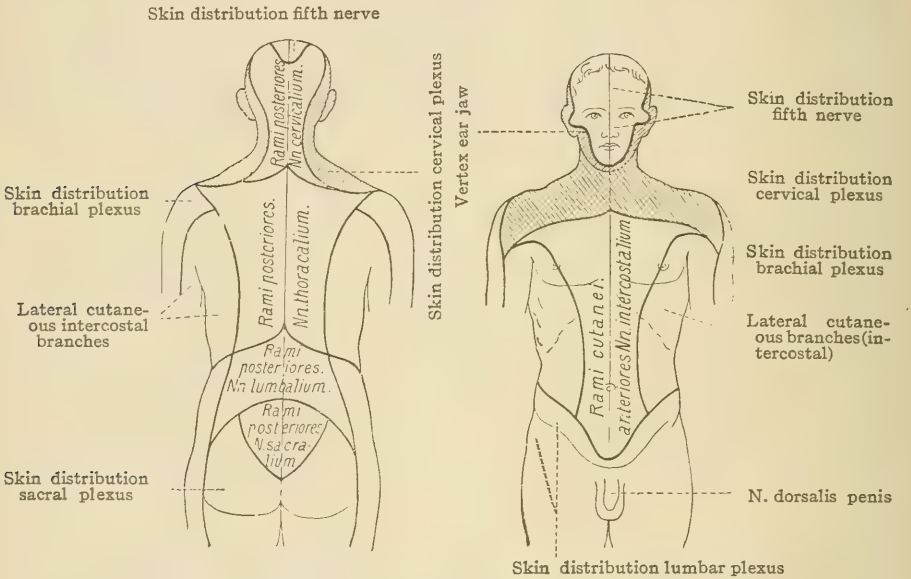


FIG. 19.—Distribution of the cutaneous nerves in the upper extremities. (*After Toldt.*)

touch in itself is always very light, but a fine gradation of the pressure of the brush touch is almost an impossibility, and yet there occurs often the necessity for ascertaining even very fine distinctions in touch sensibility. The test of the "touch circles" of the skin by simultaneously placing upon it the points of a compass at varying distances—which answers at the same time for a test of "*orientation*"—the patient to state whether he feels one or two points of contact, when the points of the compass are at a definite distance from each other, may also be dispensed with, since even upon the normal skin it gives widely varying results.

It is best to carefully touch first, parts of the body which have a *good* sense of touch, only then those that have less sensibility, and let the patient respond to every touch he feels with the word "now." But here it is necessary occasionally to touch also distant parts of the skin, in order to *avoid* a sequence of symmetrical points on extremities (which the patient himself for the most part unconsciously compares) and a definite rhythm of the touches, since through all these procedures the task of the patient is made too easy, his attention too little occupied, and I recommend also asking from time to time "Do you feel this? this? this?"—occasionally asking the question without really touching the skin. Nothing else so sharpens the attention. If one has



FIGS. 20 and 21.—Distribution of the cutaneous nerves in the trunk. (*After Toldt.*)

found parts with lessened sensibility to touch, one should try to make a boundary between these and those that have the normal sensibility and immediately observe whether this boundary is of a segmentary nature, following the distribution of the individual cutaneous nerves, or of a hysteric (glove- and sleeve-like, unilateral, etc.) character. One should note also whether the touch which is felt everywhere may not be in some regions (affected with paræsthesia) felt "different" than in those with normal sensations, which is not rarely the case. For these cases and for the lightest objective disturbances in sensibility, it is also recommended to let the patient decide whether he is being touched with the "point" or the "eye" (sharply or bluntly) of a needle which is not too sharp; with some practice, the disturbing factors of pain and pressure that are easily produced here may be avoided.

The mucous membranes which, on the whole, show somewhat slighter sensibility than the skin, may be tested in like manner, but the results are then less reliable (cf. below).

The normal sensibility of the skin varies in delicacy in the different parts of the body, being finest at the ends of the fingers and the tip of the tongue, and dullest (even to anæsthesia) upon the callous portions of the soles of the feet.

If we have examined the touch sensibility and noted the disturbance that we may have found upon a diagram of the body (which is earnestly recommended), we proceed to test the *temperature sense* or more correctly the sensations of heat and cold. For this we use test tubes, filled with warm and

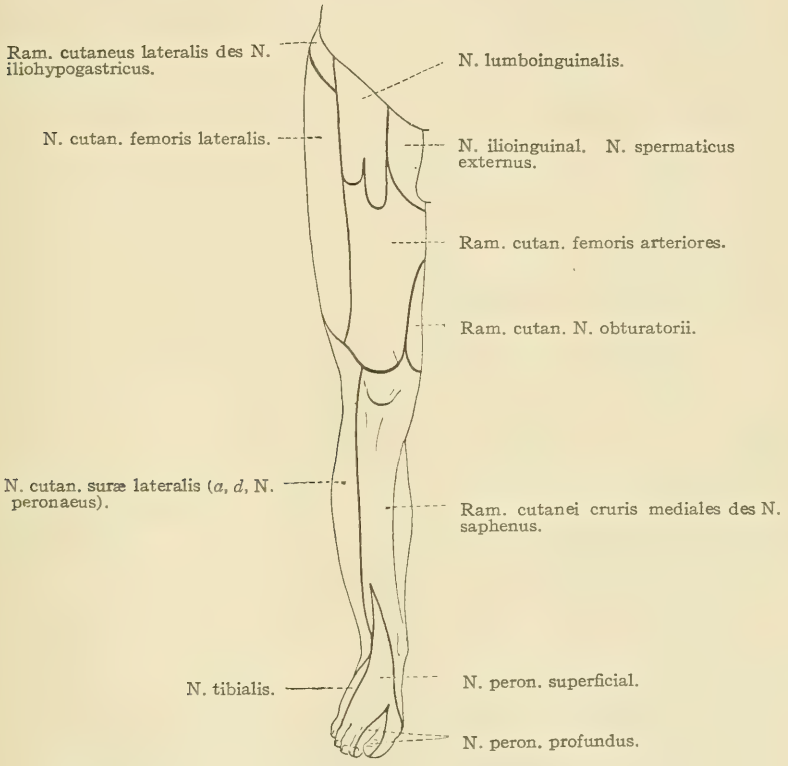


FIG. 22.—Distribution of the cutaneous nerves in the lower extremities. (*After Toldt.*)

with cold water (no extreme degrees of heat and cold, through which pain sensations are evoked), possibly also for a quick test the warm hand and the cold metal of the percussion hammer, and proceed in the same manner as described above. As a rule, the disturbances thus established will coincide more or less with those of the touch sense, though not exactly at the boundaries. This is partly due to the fact that, as *Goldscheider* has recently established on the basis of careful investigations, the skin has special points of pressure, cold and warmth, that is, places (and especially nerve ends), where touch, cold or warmth, may be best felt. But as *Oppenheim* pointedly remarks, it would be very difficult clinically to proceed to investigate the

surface of the skin, as laid out by *Goldscheider* according to this scheme, as to areas possessing varying degrees of sensibility, especially as this procedure scarcely ever gives more important results than are obtained by the usual test.

In another respect also, I recommend a practical deviation from *Goldscheider's* proposals, namely, in the test of *pain sensations*. Granted, that special points of pain and, therefore, pain-nerves too, can not be found in the skin, and that, on the other hand, by increasing almost every stimulation of any sensory quality, the sensation of pain may be evoked, still, clinically, we can not dispense with the concept of pain sensation—first because of the patient's psyche, usually very susceptible to the thought of pain (though,

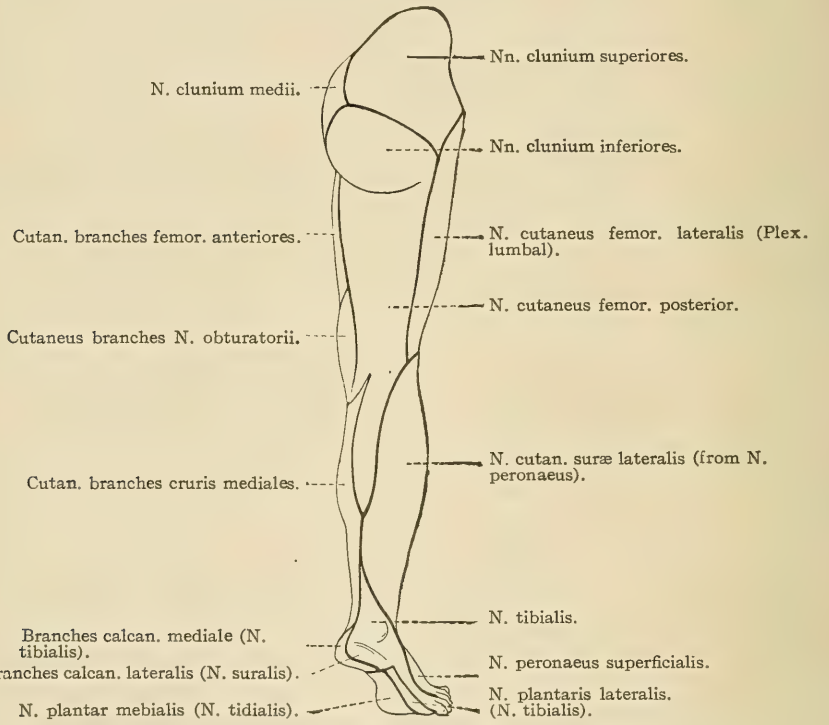


FIG. 23.—Distribution of the cutaneous nerves in the lower extremities. (*After Toldt.*)

doubtless loss of the pain sensation is often observed only late), secondly, because frequently in sensibility disturbances of functional and even of organic nature, only these more intensive, pain-producing stimulations show functional changes. It is best to test the pain sensation simply with a sharp needle and by pinching folds of raised skin, or, further, by the electric current, making use of fine electrodes. The needle point is also the principle made use of in the numerously used apparatus for measuring pain (algesimeters), in which a sharp needle may be screwed out for a greater or lesser distance from a flat surface upon the instrument; when the needle protrudes only slightly, so as not to pierce the epidermis, the application of the instru-

ment is felt only as a touch, gradually, with the lengthening of the needle, as pain. All algesimeters may be regarded as useless for clinical service.

Whether the *psycho-galvanic reflex phenomenon* described by *Veraguth* (a galvanometer inserted into the circuit of bodily conduction shows deflections in psychic processes, therefore also in pains, no beat is visible in the stimulation of analgesic zones) will be able to fill the gap, seems for the present, very doubtful; but, not having personal experience, I shall, for the time being, refrain from pronouncing judgment. For certain zones of the body there are moreover useful reflex phenomena for judging the pain sensation, as, for instance, dilatation of the pupils which occurs after painful stimulation of the skin of the cheek, which is non-existent when the cheek is analgesic.

The sensitiveness of the skin to pain is not constant over the entire body, being greatest in the skin of the forehead, least in the region of the buttocks. At every test of the pain sense, one must watch, besides the intensity of the sensation, also the period of latency between the touch and the sensation, since in no other sensory quality does such considerable "retardation" occur (cf. below). The test may, in general (except particular cases, cf. below), in order to spare the patient, be confined to those parts of the body in which one of the previous examinations demonstrated the presence of a disturbance.

The test of the sense of locality, that is, the keenness in the recognition of the places upon the skin that are touched, may without trouble be combined with the testing of the sense of touch, by having the patient, in the more delicate tests, say where he was touched. As a rule, the results do not differ essentially from those obtained by testing the sense of touch. (The test by means of the compass is in a strict sense—cf. above—also a test of the localization sense.) To the examinations of the actual sensibility of the *skin* there belongs, furthermore, according to some authors, the *hair sensibility*, that is, the sensitiveness to a slight touch upon the hair alone, not upon the skin beneath it. The areas of most delicate hair sensation are by no means identical with those of the most delicate skin sensation (but the supposition that the skin underlying the hairy parts of the body is always less sensitive, is not justifiable). The method has not led as yet to practically important results. The examination of *farado-cutaneous sensibility*, occasionally used in testing the sense of touch, that is, the feeling of tickling (formication) to weak faradic currents, seems to give results identical with those of touch and pain sensation, so that this finely graded and measurable method (determination of the stimulation threshold of sensation by means of the gradation of the current) offers no special advantages. The assertion that certain poisons (bisulphide of carbon) mainly disturb farado-cutaneous sensibility, has not been established.

Far more important are investigations of the *deep sensibility* (bones,

joints, muscles). According to *Goldscheider*, instead of the expression formerly used "*muscular sense*" which comprises the main mass of deep sensibility, one should use its separate components: the sensation of active and passive movements, the sensation of weight and resistance (also called strength sensation) and perception of situation. *Stereognosis* (recognition of the form and other external qualities of objects by means of touch) again embraces several of these qualities in another grouping, as well as the most delicate psychic processes (mnemic images). As an independent sensation, *bone sensibility* would have to be added to this. All these forms of sensibility may be disturbed individually, and in groups, with and without participation of the skin sensation.

The *sensation of active movements* may be tested in the patient only with great difficulty, if it occurs in isolation. When he is told to perform certain movements and then to state the effect of movement, the situation into which the limbs were brought, the sensation of position is virtually tested (cf. below). Unquestionably, there exist in a sound person sensations of active movement, the loss of which is felt as a disturbance (cf. below); but an actual test can scarcely be performed. The test of the *sensation for passive movements* is, on the other hand, very easy. With one hand, the examiner fixes one of the patient's extremities, and with the other, he performs as slight excursions as are possible in small joints in the peripheral parts of the extremities, small movements of the toes, excursions of the fingers, etc. The patient, who naturally must have his eyes closed or bandaged, states the direction of the movement. Tests with *considerable* excursion in the joints concerned should not be made, because through tension of the skin, tendons, etc., the patient is supplied with other possible sources of knowledge. For the same reason, the test for the large joints (knee, hip) can not be used extensively because of the large areas of skin concerned. The *sensation of weight and resistance*, the *weight sense*, may be tested in intelligent patients, by allowing them to raise, with their eyes closed, loads of various weight (with the entire extremity, or with individual parts of it, forearm, finger, etc.), and to tell the difference in weight. As a rule, this sensation is disturbed only in combination with others; moreover, the poor valuation ability of most patients destroys its usefulness, which otherwise, for instance, might easily be shaded more minutely by the dynamometer, upon which one lets the patient exert certain predetermined degrees of pressure.

The *feeling* of posture, on the other hand, permits of more exact examination. One places the extremity (or part of it) in a position contrary to that of the natural posture in rest, and has the patient state what the new position is like, about how far it is removed from the position of rest, etc.

The *stereognostic sensation* may also be very easily tested; since this is but very slightly developed without actual touch, it is best to put the object to be tested directly in the hand of the patient, and to let him describe its

form, size and substance. For this we choose as varied objects as possible, which are, however, known by their form (coins, keys, dice, balls, fabrics like velvet, silk, etc.). As has been observed above, several kinds of sensation are involved in this recognition.

Déjérine and Egger designated as *bone sensation*, the sensation of the vibration produced by a vibrating tuning fork when placed upon a part of the body where the bone is covered only by the skin. They consider it an independent sensation, because it may be disturbed without the tactile disturbance of the skin, and vice versa. This fact is probably quite correct, as German authors were able to prove also in the subsequent test (partly, however, it is designated pallæsthesia, and ascribed not only to the bones but also to all other subcutaneous tissue layers); but since this *vibration feeling* is especially common with simultaneous disturbances of the "muscular sense" (tabes), a particularly far reaching result could scarcely be expected, and has not yet been attained.

Our knowledge as to the *sensibility of the internal organs* in the narrower sense, is by no means good. The *mucous membranes* of the cavities communicating with the air (mouth, nose, cornea) show the same, though in general somewhat lowered sensibility, as that of the external skin, as we have mentioned before. This is not true of the pleural and peritoneal cavities, the lungs, stomach and intestines, the bladder and kidneys. Among all these formations, the pleura, peritoneum and the mucous membrane of the bladder are those only that are certainly sensitive to pain and touch. The meninges, substance of the brain, lungs, stomach, intestines, on the other hand, are not sensitive to touch, but may very easily be the seat of spontaneous pain sensation, which is probably induced by the sympathetic (*L. R. Müller*). But for a test of these functions, we have as yet no sufficient experience.

Disturbances in sensibility are divided first into subjective—paræsthesias and pains—and objective; namely, hyperæsthesia, hypæsthesia, and anæsthesia.

Paræsthesia signifies the abnormal sensations of the various qualities which are present only *subjectively*. They belong most frequently to the realm of the touch sensation, more rarely to the temperature sensations; the abnormal subjective sensations from the group of the "muscular sense" (cf. above) are rare; but frequently a tabetic may feel disturbances in the sensations of position, movement, etc., that can not be objectively demonstrated (he does not know how he holds the foot in walking, etc.; but here, as a rule, paræsthesia and objectively demonstrable disturbances coincide), also in psycho-neuroses, there occur occasionally disturbances of feeling of posture, of stereognostic recognition, etc., in purely subjective form. The most common paræsthesia is *tickling* (feeling of deadness, of numbness, formication, etc.), which very frequently represents an early symptom of

severe organic diseases, and moreover is often in so far objectively demonstrable that the patient feels exactly a touch in the paræsthetic area, but "differently" than in the sound parts of the body. Disturbances in heat sensation are usually described in very characteristic manner: "I feel as if my legs were packed in ice" (myelitis). A common paræsthesia, which is often combined with pains, is the "girdle feeling"; as if a hoop were placed around the body, a cord about the knee (usually corresponding to an affection of the posterior roots). In many paræsthesias, their purely psychogenous origin may be recognized at once (for instance a feeling of a living animal in the stomach, globus).[1]

Pains represent the most difficult subject matter of neurologic (often, too, of internal) diagnostics. Only in extremely rare cases can the spontaneous pain be recognized by the examiner (occasionally through vaso motor symptoms, reddening of the face, pallor, fluctuations in blood pressure, local coolness or warmth)—most often in pains evoked through the sympathetic paths (vascular crises, etc.), in which we may find conditions of collapse, superinduced by the pain, which, however, occur also in purely "sensory" pains. Unfortunately, we have no means for measuring the spontaneous pain (cf. above). In every case, we let the patient describe his pains as accurately as possible; their localization (following nerve trunks, diffuse, corresponding to an internal organ—cf. below—confined to one vascular realm), their character ("nagging," "boring," "pressing," the lightning-like "lancinating pains," "colic-like" pains); their duration may often be of great significance.

Of the *objective disturbances in sensibility*, the *hyperæsthesias* are comparatively the rarest. Tactile hyperæsthesia, the feeling of an unpleasant sensation of pain upon the slightest touch, is found occasionally in neuralgias, but also in pure neuroses and in diseases of the internal organs in the so-called "zones of Head" (cf. below). Hyperæsthesia to cold and (far more rarely) to heat is, though not a common, still a characteristic symptom of organic diseases (tabes). Hyperæsthesia to pain or hyperalgesia, that is, feeling of a violent pain upon a touch that usually causes but a disagreeable sensation or a slight pain, is, on the other hand, found rather frequently in all sorts of organic (pressure points in neuralgias of this kind) and psychogenous (*ovaralgia* in hysteria) diseases.

The group of *hypæsthesias* or *anæsthesias*, the partial or total loss of the various qualities of sensation (the single forms are known also as thermoanæsthesia, hypalgesia, asterognosia, pallanæsthesia), is the largest. This is the point in which the test of the separate sensory qualities has the most valuable results. We must first, as has been stated above, consider the distribution of the zones of such sensory disturbance (Fig. 24). Here we

[1] On the other hand, the paræsthesias of the sense nerves—entoptic and entotic phenomena—are also frequently found in non-psychogenous diseases.

find four main forms of distribution of these disturbances: types of total break in conduction in the central organ (sensory hemiplegia, paraplegia—occurring from organic, as well as from psychic causes); disturbances in the area subserving the individual segments and roots of the spinal cord (cf. the segment schemata in the special pathology of the spinal cord); disturbances in the distribution area of the peripheral sensory nerves, for which the schemata subjoined will serve as points of reference; finally disturbances of psychogenous kinds ("central type"), which are restricted to none of the forms described (glove, sleeve-stocking-like anæsthesias, usually in hysteria).

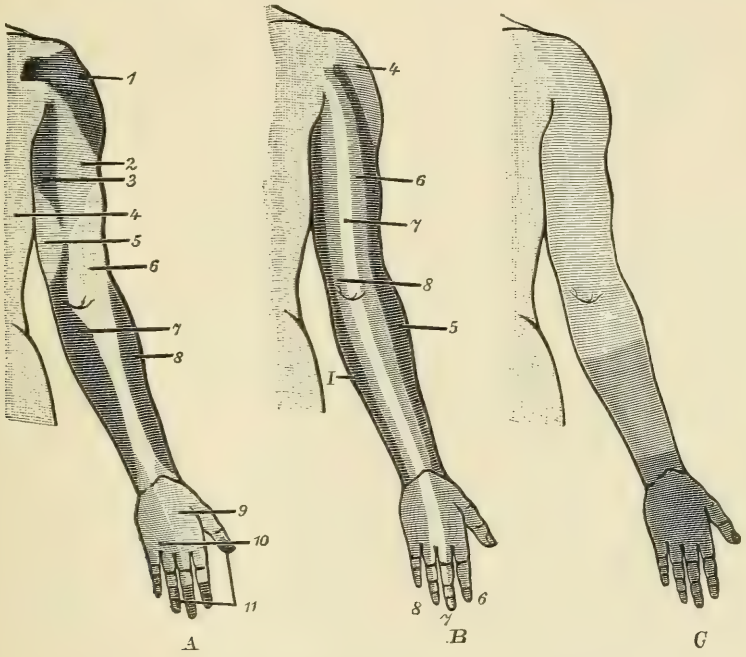


FIG. 24.—The three types of disturbances in sensibility at the extensor side of the right upper extremity. (*After Obersteiner and Redlich.*) *A*. Peripheral type. 1, N. supraclavicularis; 2, N. axillaris; 3, N. cutan. post. sup. radialis; 4, lateral branches n. spinal; 5, N. cutan. medial radialis; 6, N. cutan. post. inf. radialis; 7, N. cutan. med.; 8, N. cutan. lateralis; 9, N. radialis; 10, N. ulnaris; 11, N. medianus. (*After Bernhardt.*) *B*. Segmental type, 4, 5, 6, 7, 8. 4–8, Cervical segment; I, 1. Dorsal segment. (*After Allen Starr.*) *C*. Central type.

To which of the groups mentioned, the hypæsthesia or anæsthesia under consideration belongs—a question often decisive for diagnosis—is a matter that can be settled only by determining accurately the boundaries of the disturbance. Inasmuch as an anæsthetic area but rarely touches directly one that has normal sensibility, this determination demands much patience and skill.

We must also mention here, that *Head* succeeded in finding for diseases of the internal organs (lungs, stomach, intestines, etc.), cutaneous nerve areas, which synchronously display abnormal forms of sensibility, according

to what has been determined so far, almost exclusively hyperæsthesias ("reflex hyperæsthesias" best known in gastric affections, as band-like or singularly bounded zones of half the abdomen, of the back, etc.). We can not here enter upon a detailed discussion of this subject.

As has already been said, each quality of sensibility may be individually disordered, or they may all be affected at the same time. We find, rather frequently, also, an affection by groups; *only* the cutaneous, or *only* the deep sensibility. The most common of these forms of a partial or "*dissociated sensibility disturbance*" is probably that of simultaneous disturbance of the pain sensation and of the cold and heat sensation, while the tactile sensibility remains normal, as in syringomyelia.

Some rather rare disturbances shall be briefly mentioned. The pain sensation is not rarely disturbed in such a way, that the pain stimulation is felt only late, after a period of latency of one or more seconds, that is, is felt as pain; as a rule, the touch is at once registered as such, and only the pain appears later (delay or *retardation of the pain sensation;* double sensation). In other cases, a single, brief, though intense pain stimulation (prick) produces no pain; on the other hand, pain is felt, if one allows it to persist for some time with the same degree of severity (*summation*). Disturbances in the temperature sense are most frequently the foundation of "*perverse sensations*" which are as such, rare; warm is felt as cold, cold as touch or pain, etc. *Allocheiria*, the localization of touch upon the corresponding place in the extremity not touched, is, at any rate, as rare an occurrence as *polyæsthesia*, in which a single touch is felt as if multiple.

The characteristic disturbance of *deep sensibility*, frequently met in central organic nervous diseases, affects most frequently the sense of position and the stereognostic sensation. The patient in bed "no longer knows where his legs are, whether he has any or not"; he no longer recognizes objects placed in his hands. Disturbances in the sensation of vibration are decidedly less common.

E. Reflexes

We distinguish *tendon reflexes*, *cutaneous* (and mucous membrane) *reflexes and internal reflexes*.

Without entering with more detail into the concept of "reflex" we shall mention here only that we shall designate as "reflex" in the sense of the phenomena now under consideration, only muscular twitchings, that is, contractions, which follow a sensory stimulation, with the exception of the idiomuscular contractions (cf. above). All these reflexes, then, have a local motor effect. We regard in general, also, the tendon phenomena discovered by *Erb* and *Westphal*, as genuine reflexes, not (as *Westphal* held) as direct stimulation of the muscle concerned.

There are an immense number of *tendon reflexes* in the human body, many of which can be proved present, even in the normally healthy individual, though not all constantly, and some only upon increase of the reflex. We shall mention here only the most important, the knowledge of which quite suffices for the recognition of all nervous diseases involved.

In the head we know only the *lower jaw* or masseter reflex. It is most easily evoked as follows: When the mouth of the patient has been slightly opened the examiner places a finger upon his lower jaw and with the percussion hammer, gives the finger a light blow from above; there results a raising of the lower jaw, effected mainly by the masseters, the mouth closing. The same reflex may be evoked by the periosteum of the lower jaw near the point of insertion of the masseters; this reflex is virtually one of the so-called periosteal reflexes, which, however, genetically and nosologically, may be counted among the tendon reflexes. The lower jaw reflex is comparatively constant in the healthy.

The somewhat less constant *forearm reflexes* are also genuine periosteal reflexes and may be evoked by tapping the lower end of the radius (radial periosteal reflex) and the ulna (ulnar periosteal reflex) which causes, in the (more common) radial reflex, a reflex contraction of the supinator, that is, a flexion of the forearm, in the ulnar reflex, usually a slight retardation. Both reflexes are tested with the forearm flexed into a right angle, adducted and slightly pronated, in order to relax the m. supinator as far as possible.

The *triceps reflex* is scarcely ever lacking in the healthy, but is often difficult to test. The arm of the patient is held somewhat in the same flexed position as mentioned before, and a short, quick blow with the percussion hammer, delivered upon the tendon of the m. triceps, right above the olecranon. There results a movement of extension, often only slight, in the forearm. The danger consists in the possibility of striking the muscle itself instead of the tendon, thus producing direct muscular stimulation, which, as a rule, has a slighter motor effect, but may otherwise closely resemble the reflex. Since the triceps reflex—a genuine tendon reflex—may be of diagnostic importance (especially its absence in tabes, etc.), this point must not be overlooked.

The tendon reflexes in the trunk are of no great importance. We must mention the *scapulo-humeral reflex* described by *Bechterew*, a periosteal reflex produced by tapping the scapula, which, however, I have not found sufficiently constant to attach great weight to its absence.

The most important tendon reflexes belong to the lower extremities. Important above all is the *patellar* (kneecap) *reflex*, the contraction of the m. quadriceps upon tapping the patellar tendon below the patella. It is the most constant of all reflexes in the healthy (lacking only in 0.04 % of the cases) and can be tested with comparative ease. For its test, still more than

for that of all other reflexes the principle that to evoke the reflex the muscle concerned—here the quadriceps—must be totally relaxed, holds good.

Passive relaxation is usually attained by a certain position of the leg; but often even in intelligent, especially in timorous, patients an active relaxation is accomplished with the greatest difficulty. We recommend the following ways and means:

1. The patient sits on a chair, crosses the leg to be tested over the other; the examiner delivers a horizontal blow with the inner edge of his hand or with the percussion hammer upon the quadriceps tendon below the patella, where it bridges over the space between the patella and tuberositas tibiæ and lies therefore in a more or less hollow space (this "hollow space" is probably the reason for its remarkable constancy). This method is not safe because total relaxation is rarely attained; it has the advantage of the largest motor effect (of the sudden lifting of the tip of the foot) which, however, is of comparatively less importance because in excitation of the patellar reflex one should always pay more attention to the contracting muscle than to the motor effect; in decreased reflexes, it often happens that the former only is visible.

2. The patient sits on a chair which is to be several centimeters lower than the leg proper of the patient and places the foot of the leg to be tested so far forward that leg and thigh form an angle of about 100 degrees, and the thigh rests easily upon the chair. A great many patients are best able to relax the muscle when in this position; the motor effect is slight, but the contraction of the muscle is usually very distinct. I recommend this method, as well as the following:

3. The patient lies in bed, as flat as possible, with the upper part of his body but slightly raised; the leg is somewhat flexed so that leg and thigh form an angle of about 140 degrees and at the same time the thigh is abducted (the knee of the flexed leg inclines to the outside). The blow on the patellar tendon usually evokes an evident extension of the leg proper. The relaxation of the quadriceps usually succeeds, though not always easily. Crossing of the legs in a recumbent position is not so good.

4. The leg proper of the recumbent patient is raised by means of a handkerchief or some similar device placed beneath it, so that the leg lies horizontally in its support and the thigh is flexed into obtuse angles with it and the trunk. Here, it is supposed, the relaxation is very nearly complete. Almost every year, however, new modifications are recommended for testing this reflex, that is, for relaxing the muscle.

I should call the *Achilles tendon reflex*, considering its work and significance, the second of the most important tendon reflexes of the body. It consists in a contraction of the calf-muscles from which the tendon of Achilles courses, of the triceps suræ, and thereby evokes plantar flexion of the foot. It is elicited by striking the tendon of Achilles with the percussion hammer; here, too, the relaxation of the calf is the main thing. For this, the position

described above under 3 usually suffices; the examiner then stands at the foot of the bed and from the inner side of the leg delivers a brief blow upon the tendon. Under certain circumstances one must test the tendon at several places. The absence of this reflex, as that of the patellar reflex is, in every case, pathological (unless contractures, etc., prevent their appearance). Other methods for testing it are the striking of the tendon while the patient kneels on a chair, or by striking the tendon while the patient lies on his stomach with his leg flexed at the knee.

All other tendon reflexes are inconstant, that is, demonstrable only in pathological increase of the reflex, even the *dorsal foot reflex* described by *Bechterew* and *Mendel* (which with *Spier* I would count as a genuine tendon phenomenon.)

We know two kinds of pathological changes in the tendon reflexes: *increase* and *decrease*, or *absence*. The former condition is accompanied by increase of the muscular tonus (cf. above), being its most regular concomitant phenomenon, the latter with decrease of the tonus (but here the connection is less constant).

We can say with certainty when a reflex is to be called *increased* only in the case of the patellar and the Achilles tendon reflexes. Increase in the reflex, as a rule, effects a more extensive muscular contraction with considerable motor effect and also the appearance of tendon reflexes that are not demonstrable at other times. In spastic conditions of high degree (i. e., naturally only so long as the spastic musculature is not *so* tightly contracted as to make further contraction impossible) by striking almost any tendon in the body that is to some extent exposed, one may produce twitchings in the muscle concerned; among the most common in this case are twitchings in the adductor tendons of the thigh (adductor reflex) and in the tibialis posticus.

Increase in the patellar reflex is present if the reflex can be evoked not only at the tendon above and below the patella and from the patella itself, but also by striking the periosteum of the tibia in a more or less extended area. In some cases where the increased patellar reflex is evoked, the quadriceps of the other leg contracts also (crossed reflex). The highest degree of increase is called *patellar clonus;* here the quadriceps is affected by more or less rapid contractions, which last for some seconds, when, after the muscle has been relaxed as far as possible, one seizes the patella firmly with two fingers and moves it suddenly towards the tibia.

Foot-clonus (ankle clonus) as an expression of the *increase of the Achilles tendon reflex* can be more constantly evoked. When the legs are totally relaxed (position as above under 3) the examiner with his right hand placed under the sole of the patient's foot gives a sudden jerk upward towards the sole of the foot, while simultaneously the left hand protects and slightly fixates the leg proper at the knee joint. The calf shows slow or more rapid clonic contractions accompanied every time by twitching movement of the

foot downwards. It is important here that the *pressure* of the hand upon the sole of the foot must be relaxed, as soon as the blow at the sole of the foot has been delivered; the hand then remains but lightly *resting* against it (but may not let the sole go). As a rule, 10 to 20 such twitchings appear, decreasing gradually; in severe cases, however, the *foot-clonus* may persist for minutes. From this must be differentiated the *pseudoclonus* of hysterics, in which either the calf comes more nearly into a state of tremor or is even actively contracted, or some—about 3 to 4—clonic twitchings appear as expression of a slight "reflex hyperæsthesia." *Hand-clonus*, clonic flexion movements in the flexors of the hand and fingers, which may be evoked as the expression of a high degree of increase in the reflexes in the upper extremities by a sudden jerk against the relaxed metacarpus, from below upwards, with fixated forearm, is rarer.

Increase of the tendon reflexes is found, as has been stated, almost constantly in increase of the muscular tonus (cf. this) therefore in all so-called spastic diseases (the highest degrees of it in diseases of the pyramidal tracts), besides this, also in neurasthenia and hysteria; but here, as a rule, the criterion of a genuine "clonus" is lacking.

The *decrease* of the tendon reflexes is difficult to judge. We may say that we can speak certainly of a decrease in a (otherwise constant) tendon reflex when it is evoked but feebly even with the assistance of *Jendrassik's manœuvre* (reinforcement method), or also, when there is an obvious difference between the similar reflexes of each half of the body and the stronger reflex must be regarded as normal, not increased. By *Jendrassik's* reinforcement method we designate the strong active tension on the part of the patient of a muscular area, during the test of the tendon reflexes of another area. Usually the patellar reflex is concerned. If presence of the reflex seems questionable one has the patient exert tension upon the musculature of his arms, by pulling apart with all his might his hands tightly interlocked, without letting them go (or by having the patient heartily grip his own hand). At this moment the exciting blow upon the tendon is delivered. Whether in this process it is a matter of "paving the way" for the reflex, and not of merely distracting the attention of the patient and thereby securing better relaxation of the muscle, is not yet known.

If even with this help the reflex can not be evoked, we describe it as *absent*. The absence of the patellar reflex (and almost as certainly that of the Achilles tendon and triceps reflex) are found in diseases with lowering of the muscular tonus (tabes), in neuritis, in narcosis and coma, in muscular atrophies of high degree, in rare cases also, apparently congenital. Occasionally there occurs "exhaustion" of the tendon reflexes; after long testing they become weaker and more difficult to evoke.

Among the *cutaneous reflexes* only three are of great importance; the plantar, the cremasteric, and the abdominal.

We designate by *plantar reflex* a movement of the toes, which occasionally extends to the metatarsus, upon irritation of the skin of the sole of the foot (done best with the handle of the percussion hammer, also with pieces of ice, by pinpricks, etc.). The stimulation should be performed quickly and energetically; if the patients are anxious it is best to fixate the leg proper. Sometimes the reflex of certain parts of the planta pedis can be evoked with special ease (usually this is done best from the inside edge).

The *normal plantar reflex* consists in an energetic plantar flexion of all the toes, occasionally with simultaneous dorsal flexion of the metatarsus, or of the entire foot (flight reflex).

The reflex is not absolutely constant in the healthy. It is absent in every severe hypæsthesia of the planta, also in severe cornification, edema, cold; furthermore in sleep and in narcosis—besides in some organic diseases, neuritis, cerebral and spinal paralyses. Its mere absence—as well as its increase—is but rarely of great diagnostic importance.

Under certain conditions we find, however, instead of the normal plantar reflex, an abnormal reflex named after *Babinski:* the so-called *Babinski toe reflex.* Here there appears upon irritation of the skin of the planta a *slow dorsal flexion* of the large toe, occasionally, too, of the other toes and the middle part of the foot instead of the normal, rapid plantar flexion. Here we must emphasize the fact, that one uses at first but weak stimuli, slight strokes of the percussion hammer, etc., and stimulates various parts of the planta, one after the other. If this is done too energetically, there is often a rapid withdrawal of the foot and an undecided wavering of the toes, composed of extension and flexion, which can be interpreted only as flight reflex. The *Babinski* phenomenon is found normally only in sucklings. In later life it may be regarded as a pathognomonic sign of disease of the pyramidal tracts. As to its origin, whether it is only a modification of the normal plantar reflex or is an independent reflex, opinions are still divided; it is probably a matter of a purely spinal reflex, which can appear only after suppression of the normal plantar reflex which goes through the cortex of the brain.

Oppenheim's phenomenon may be described as to a certain degree analogous to the *Babinski* phenomenon. Upon *energetic* stroking of the skin of the leg at the inner edge of the tibia, there is, in the healthy person, no reflex movement of plantar flexion of the toes; in spastic conditions, on the other hand, there appears a dorsal flexion of the foot and the toes. I found the phenomenon less constant than the *Babinski*, with which it generally appears, and its excitation frequently somewhat irritating to the patient. In the same group belongs also *Remak's* femoral reflex: upon stroking the inner side of the skin of the thigh, there results reflex raising of the entire leg and dorsal flexion of the foot—usually only in pyramidal tract lesions of the spinal cord. Probably in all these "pyramidal tract"

reflexes, a synergism of certain groups of muscles (extensors of the foot and toes, flexors of the leg proper, ileopsoas) appearing only in pathological cases, plays the main rôle; it may be tested in another way, too (*Strümpell's* tibialis phenomenon): the patient is asked to draw his thigh energetically up to his trunk while the examiner exerts strong resisting pressure upon the thigh; in certain diseases (according to *Strümpell* especially in multiple sclerosis) the synergism mentioned appears especially in an energetic contraction of the m. tibialis anticus.

Furthermore there occur occasionally, also in mere increase of the usual plantar reflex, co-movements in the musculature of the thigh of the same, sometimes, too, of the other extremity and even plantar reflex on the side that has not been stimulated.

The *cremasteric reflex* consists in a prompt contraction of the cremaster muscle, that is, an elevation of the testicle on the side in question, occasionally too on the opposite side, upon stroking (prick, cold) the inner aspect of the thigh near the scrotum. The reflex which in the female is replaced by the analogous inguinal reflex and is rather constant, is important only when there is some difference between the two sides and in total absence, especially in transverse diseases of the spinal cord, in which the knowledge of the location of a reflex arc is of importance for exact local diagnosis.

The same is true of the *abdominal reflexes*. We know of three such on each side—the upper or epigastric, the middle and the lower (hypogastric) reflex. They are evoked by transverse stroking of the upper, middle, or lower third of the skin of the abdomen and consist in a sudden contraction of the obliqui and the transversus abd. In most cases they can be evoked separately; but frequently they are altogether lacking when the abdominal walls are very flaccid or very thick (fatty, edematous). Their absence, moreover, may be considered an early symptom of multiple sclerosis. The absence of separate abdominal reflexes may attain great importance for localization in myelitic processes.

We must mention briefly the almost constant *anal reflex* (contraction of the sphincter ext. upon pricking or a similar irritation of the skin of the anus) and the *scrotal reflex*, the peculiar slow contraction of the tunica dartos of the scrotum, which upon stroking or refrigeration (uncovering) passes in curious waves over the scrotum. The latter is remarkable because at least one of these reflex paths might pass through sympathetic fibres. These two reflexes have not yet attained any clinical importance worth mentioning.

In respect to the *mucous membrane reflexes* we can be very brief. Only the *corneal or eyelid* and the *vomiting or retching reflex* are important. In the eyelid reflex there appears upon touching the conjunctiva or cornea immediate contraction of the orbicularis (to be distinguished from the reflex closure of the orbicularis upon touching the lashes, which is probably to be considered

a cutaneous reflex), and the eye closes. The vomiting reflex consists of a retching movement produced reflexly by touching the posterior wall of the pharynx. Both reflexes are occasionally found to be changed (lacking) in organic processes in the brain and the medulla oblongata. But besides this, they are frequently lacking in psychogenous affections (hysteria); but since they are not constant even in the normal individual and (especially the vomiting reflex) can be suppressed to a great extent by will-power, the title of hysterical stigma has been unjustly assigned to them.

Of the *internal reflexes* we have already discussed one, the pupillary. But we must mention here, furthermore, the reflex processes subserving the *evacuation of feces and urine* as well as those of the *sexual sphere*. It is true that we have no real test for these reflexes; but the pertinent statements of the patients and the clinical observation of the physiological course of these processes permit of a rather accurate representation and the leading sphincter disturbances ought not to be neglected in any general nerve examination.

Evacuation of urine and feces is indeed subserved by both active and reflex processes. The *strong desire* to evacuate, which occasionally even in the normal individual overcomes the resistances of the sphincters, is certainly for the most part reflex, caused by the degree of fulness in the intestines or the bladder or by the manner of filling (irritation of the walls by pathologically changed contents). The disturbances of these functions are called *incontinentia* alvi (urinæ), that is, the inability to hold back the contents, and *retentio* urinæ (retentio alvi), the impossibility of active evacuation. Naturally these names are also used promiscuously for the disturbances of the active powers concerned for voluntary evacuation. Still we may regard the retention phenomena, such as are found in cerebral and high lesions of the spinal cord, as well as incontinence in destructions in the lower segments of the spinal cord, with some justification, as a disturbance of the reflex of evacuation of urine and feces.

The *reflexes of the sexual sphere* are only of slight importance for the diagnosis of organic diseases, with the exception possibly of the phenomenon of impotence (cœundi) in tabes and possibly as a rare symptom of the phenomena of functional loss in diseases of the lowest segments of the spinal cord, analogous with the animal experiments of *L. R. Müller* (loss of ejaculation in disturbances of the sacral portion of the cord). All the greater is the importance of the numerous psychogenous disturbances which belong to this realm, such as psychic impotence, priapism, too frequent pollutions, etc. Especially in the various forms of neurasthenia (cf. in the special part) do we meet those reflex disturbances, to which strictly speaking, many disturbances in menstruation also belong. It is common to all internal reflexes, with the exception of the pupillary alone, that the paths of the sympathetic nervous system play an important rôle in producing them.

F. Vasomotor, Trophic, and Secretory Disturbances

To this chapter belong the least investigated disturbances of the entire neurological field. This is true especially of *vasomotor* phenomena of the origin of which we know only that they possess centers in the medulla oblongata and their paths for the most part or altogether are the sympathetic tracts; disturbances of this kind are found in neuroses (partly independent) and in diseases of the cervical sympathetic, in the latter case, in a definite arrangement. As concomitant symptoms of organic diseases they are rarer.

Abnormal redness or *pallor* of the skin, abnormal *warmth* or *cold* are naturally to be considered disturbances only when they follow inadequate stimuli. Occasionally such a condition of abnormal redness, pallor, coolness, persists for days and weeks, apparently caused by a paralysis or a spasm of the vaso-constrictors. The most typical form of such local vascular disturbances is *Raynaud's disease* (symmetrical gangrene), the incipient stages of which are mostly characterized by pallor and coldness of the end phalanges which later passes into gangrene. Similar conditions of temporary nature play the main part in *intermittent claudication* (dysbasia or claudicatio intermittens arteriosclerotica), where through vascular spasm, pallor and coldness of the skin, pains and disturbances in movement appear. From this one must differentiate the *akinesia algera* (*Möbius*), general motility disturbance upon a foundation of psychogenous pains. Also in abnormal redness due to vascular paralysis (or dilator spasm) pains may appear in rare cases (*erythromelalgia*). Occasionally these vascular disturbances are unilateral, for instance, in the face in disease of the cervical sympathetic.

Dermographism (urticaria factitia) is to be understood as abnormal irritability of the vasomotors, as it is found in some neuroses and especially in meningitis; upon rapid stroking of the skin (finger nail, percussion hammer) there appears upon the spot stroked a temporary reddening, then an anæmic white swelling which persists for minutes and even hours, possibly caused by exudation into the tissue.

These serous exudations often appear in the form of local or general edemas which are distinguished from those in renal disease only by the normal findings in the circulatory apparatus, as well as by their transitory nature; in older cases also by their great obstinacy. There has been described as *intermittent edema* an independent "vasomotor" neurosis in which a local edema appears now here, now there, in the skin or mucous membranes, lasting some hours or even days. The joints also may be affected by similar disturbances (*hydrops articular. intermittens*). In hysterics we distinguish the so-called blue edema (with vascular congestion) and white edema (with anæmia) which mostly appear locally, unilaterally, and may be very obstinate.

The *trophic disturbances* appear in more striking form than the vasomotor. Besides the muscular atrophy already discussed, the skin and skeleton frequently exhibit trophic disturbances dependent upon nervous diseases. Some form a borderland between neurology and dermatology. Among these I reckon *scleroderma*, in which circumscribed or diffuse parts of the skin (especially forehead, nose, fingers) through loss of elasticity and shrinking of the skin become smooth, glossy, usually brownish in color; the skin can no longer be raised from the parts beneath and considerable restrictions in movement result. Furthermore, *herpes zoster*, a blister-like eruption upon certain segments of the skin, accompanied with neuralgiform pains, *ichthyosis*, usually a congenital disease characterized by abnormal dryness and desquamation of the skin, the cutaneous atrophy known as "*glossy skin*," in spinal cord and nerve trunk disease, *myxedema*, in which besides a diffuse swelling and pallor of the skin (though upon pitting the depressions do not persist), trophic disturbances in the nails, hair, etc., may also appear. Trophic disturbances of the hair are by no means infrequent; loss of hair in spots (alopecia) and sudden diffuse or circumscribed canities are the main types, both of which frequently appear after or because of nervous diseases.

The most severe forms of atrophic skin affections are the *ulcers*, among which we cannot, of course, include the artefacts of hysterics who occasionally produce ulceration themselves. But there does belong here the *mal perforant* of syringomyelia and tabes, the spontaneous occurrence of deep, sharp-edged ulcers, especially on the planta pedis, and, furthermore, the already mentioned *Raynaud's disease* in its last stages. In a certain sense we must include here also the extensive *decubital sores* (due to pressure upon anæsthetic places) of myelitics and the mutilation of the phalanges in the so-called "*Morvan's* type" of syringomyelia.

Trophic disturbances in the *bones and joints* are seen in the form of atrophies as well as of hypertrophies. The former appear especially in spinal diseases accompanied by muscular atrophy, as in *infantile spinal paralysis*, where usually the bone of the extremity affected remains behind in growth as the musculature disappears; hypertrophies form the characteristic of *acromegaly*, where the thickening of the bones (especially in the forehead, chin, fingers and toes) besides the thickening of the other tissues is the substratum of the disproportionate condition of the "acra," the ends of the extremities. Destruction of bones is found in *spontaneous fractures* and especially in *arthropathies* as in severe spinal diseases (tabes, syringomyelia). Here there appear painless inflammations of the joints accompanied by hydrops and pronounced thickening, later destruction of the capsule and the articular ends of the bones, which in their turn may lead to fractures, luxations and subluxations (Fig. 25).

The *secretory disturbances* form the smallest group of this division. If

we do not include among them the above described disturbances in evacuation of the feces and urine, only abnormal conditions in *secretion of sweat* and *saliva* are concerned. We observe occasionally an abnormal dryness of the mouth in tabetics, abnormal increase in saliva (salivation, ptyalism) in epileptics, affections of the sympathetic, etc. The sweat secretion may

FIG. 25.—Arthropathy and hypotonia (genu recurvatum in tabes dorsalis). (*After Schoenborn-Krieger, Klinischer Atlas der Nervenkrankheiten.*)

occasionally be present in the form of a defect (*anhidrosis*) in the above-mentioned cutaneous affections (ichthyosis), and may occur also in some forms of neuritis and in spinal diseases. More common is *hyperhidrosis* in all possible organic (syringomyelia, neuritis) and especially in "functional" nervous diseases (neurasthenia, Basedow's disease); frequently it is unilateral, corresponding to the localization of the process and not rarely very easily influenced by the psyche.

Of all the above-mentioned disturbances of vascular innervation and of the trophic centers we can control the occurrence only of a very few. For one single group of these symptoms there are more favorable conditions, i.e., disease of the cervical sympathetic. We know clinically, in addition to the experimental results of stimulation and total division of the cervical sympathetic, that in *paralysis of the cervical sympathetic*, contraction of the palpebral fissure and the pupil on the same side, occasionally, too, constriction of the cutaneous vessels and anhidrosis on the side concerned may appear. Frankly, the findings are not altogether constant, but the opposite symptoms which would naturally be expected from irritation of the cervical sympathetic are still less constant.

G. Examination of the Cerebro-spinal Fluid

This examination, especially since the increase in epidemic meningitis, should be familiar to the practising physician and to the neurologist. It is connected with the performance of *lumbar puncture*, the inventor of which was *Quincke*.

For lumbar puncture we need as instruments a sharp-pointed canula, 10 cm. in length, of a diameter at most of 1 mm. and a suction tube connected with it by means of a rubber attachment. During its performance the patient lies on his side or sits on a chair, in each case with spinal column flexed far forward. The canula supplied with its mandril is inserted, with all antiseptic precautions, between the 2 and 3, 3 and 4, or 4 and 5 lumbar vertebræ, about 1/2 cm. laterally from the median line and in direction inward and upward (in children in the median line sagitally upward). After piercing the muscles, the instrument meets in the ligaments before and in the foramen intervertebrale a slight, then in the dura, a still slighter hindrance. Now the point of the canula is in the arachnoidal sac which is filled with liquor; the mandril is withdrawn and the liquor drips or spurts from the canula. Now the suction tube and the rubber attachment may be connected and the pressure of the fluid be measured at once, the pressure normally amounting to 40 to 130 mm. of water and pathologically may rise to several hundred millimeters. The experienced may judge the pressure as normal, decreased, or increased from the manner in which the liquor leaves the canula, whether it be a rapid or slow dripping, or a sudden spurting. In this case we take for diagnosis only 4 to 6 c. cm., though for therapeutic reasons, it is true, often far larger quantities (up to 40 to 60 c. cm. at one sitting, especially in various forms of meningitis). Then the needle is rapidly withdrawn, the small wound closed with adhesive plaster. The operation (unless, from the restlessness of the patient, the needle swerves into the periosteum) is almost painless. On the other hand, there appear frequently, especially in patients with normal fluid, a few hours after the

puncture, pains in the nuchal region and the head, which may be very violent and last for several days; they never have serious consequences and are easily overcome if the patient will lie quietly on his back. Only in tumors of the posterior cranial cavity are there occasionally after the puncture sudden displacements within the skull due to change in the pressure (closure of the aqueduct) and now and then such a patient dies; in tumors of this region, therefore, puncture should not be performed.

The *fluid in its normal condition* is as clear as water and contains only minimal traces of albumen and very few cells.

Pathological changes may affect the pressure, the chemical and microscopic condition of the cerebro-spinal fluid.

The *pressure* is rarely diminished, on the other hand, often increased (in hydrocephalus, tumors and often in meningitis) as has been stated above.

Concerning its *chemical condition*, the albuminous content is of special importance. Increase in the quantity of albumen (apparently, in particular of the serumglobulin) is found in purulent or hemorrhagic processes of the meninges and of the surface of the brain and the spinal cord, furthermore in tubercular meningitis and above all in progressive paralysis of the insane, where it may prove to be an important differential diagnostic factor; pure globulin increase seems to occur besides these also in other metasyphilitic diseases of the cerebro-spinal system.

The *microscopy of the fluid* concerns its cellular content (*cytology*) and the micro-organisms it contains. We speak of an increase of contents, especially of small uninuclear cells ("lymphocytes") if the number of these formations in the visual field, in an enlargement of 400, amounts to more than 4 to 5 in the average. This holds good especially for all metasyphilitic diseases (gummatous processes, tabes, progressive paralysis of the insane) and for meningitis of all kinds. Occasionally, too, one finds large quantities of polynuclear cells (fresh meningitis and progressive paralysis). For the purpose of this examination, a drop of the strongly centrifuged fluid is taken from the bottom of the centrifugation glass, carefully dried on the object slide, eventually fixed and stained with methylene blue or *May-Grünwald's* staining solution, avoiding all rough measures (rough washing). It is important *not to delay* the examination. Of micro-organisms there are found in meningitis forms always corresponding to the ætiology, most frequently tubercular bacilli, then meningococcus intracellularis, pneumococcus, influenza bacilli, etc. The test (in the centrifuged fluid or in fluid which has been set aside, same staining as usual) may be very troublesome. The examination of the fluid may be of decisive importance especially for meningitis and for cases of doubtful postsyphilitic diseases. In all pure psychoneuroses and neuroses (at least when no syphilis appears in the history) the fluid is normal.

Cerebral puncture described by *Neisser*, *Pollack*, *Pfeiffer* and others may

be spoken of as an extension of lumbar puncture. In this a fine drill driven by electricity is used to pierce the cranium and then fine canulæ may be driven deep into the substance of the brain through the punctured hole.

For the diagnosis of cysts and (by removal of bits of substance) of tumors, the method is unusually valuable, presenting no danger worth mentioning to the patient; for the present, however, its use should still be restricted to hospitals and clinics.

H. Examination with Röntgen Rays

Unfortunately the Röntgen process that has come to be so invaluable for internal medicine, has as yet been of little use and help in neurology. Indirectly, of course, it may be valuable, if, for instance, it proves the presence of an aortic aneurism as causation of a recurrens paralysis—of a bone fracture in a neuralgia. But for actual neurological diagnostics, one should try to learn clearly what the method *can* be expected to do; of course, it is out of question to speak of proof of columnar diseases in the spinal cord, neuritis, diffuse cerebral diseases and all neuroses. The chances in trophic disturbances are somewhat better. In arthropathy, mal perforant, *Raynaud's* disease, acromegaly, we can well establish the hypertrophies and destruction of the bones upon the radiograph. This may be possible also for muscular atrophies. Tumors of the substance of the brain or even of the spinal cord are unfortunately only very rarely or not at all to be distinguished from parts covered with osseous tissue, which give quite diffuse shadows. Only processes in the bone itself, that is, tumors of the base of the skull or the cranial cap with pressure upon or extension to the brain, as well as vertebral affections (tumors, caries) can easily be made visible—the latter alas—not nearly so frequently as is to be desired (especially in incipient cases) and only in favorable locations (lumbar vertebræ, sacrum, cervical vertebræ). Proof of the presence of foreign bodies (bullets) is always an easy and a well-repaid task of radiography, whose results deserve attention even with regard to neuro-therapeutics (*Fürnrohr*).

II

DISEASES OF THE PERIPHERAL NERVES

BY

H. STEINERT (Leipsic)

INTRODUCTORY ANATOMICAL REMARKS AND DEFINITION

The peripheral cerebro-spinal nerves consist principally of fibres of the peripheral motor and sensory neurons (teleneurons). Morphologically and genetically the nerves of smell and sight occupy a special position. Of the other cerebral and the spinal nerves it may be said that their motor fibres emerge from the large motor cells located within the central organs, from the anterior columns of the spinal cord and from the nuclei of the brain stem, which are comparable to the anterior horns of the spinal cord. The sensory fibres, on the other hand, emerge from the spinal ganglion cells, and in the domain of the cerebral nerves, from the cells of the ganglia of the head, which, likewise located outside the central organ, are closely related to the spinal ganglia in structure and development. These are the Gasserian ganglion; the spiral and vestibular ganglia of the eighth nerve; the superior and petrous ganglia of the glossopharyngeal, and the jugular and nodosum ganglia of the pneumogastric nerve.

Each *spinal nerve* originates from the spinal cord by two roots, the anterior and the posterior. Through the anterior roots the centrifugal, chiefly motor conduction, fibres emerge from the spinal cord; through the posterior roots the sensory fibres enter into the spinal cord. If the roots themselves or the spinal nerve be injured immediately after the roots coalesce, the most important clinical symptoms—those of the loss of function—will be almost identical with those resulting from an affection of the peripheral neuron within the spinal cord segment to which it belongs. These clinical symptoms are therefore treated in connection with the diseases of the spinal cord, in whose topical diagnosis they play an important part. They also play an important part in dealing with the diseases of the membranes of the spinal cord, in which the roots of the spinal nerves are imbedded, so that in the main we look upon these diseases as partial symptoms of diseases of the spinal cord membranes.

Shortly after leaving the spinal cord the most important spinal nerves undergo complicated anastomoses and interlacings of their fibres. Those

fibres belonging to the innervation areas, to the cutaneous areas and to the single muscles, no longer are to be found grouped in bundles, in which shape they were related to the central organ. Fibres of the same spinal nerve roots separate into different branches of the nerve plexus; fibres of different roots unite. In this way there arises an anatomical basis for the appearance of new and distinct clinical pictures. The symptoms after lesion of certain trunks of the plexus are grouped in a characteristic way, suited to the new anatomical conditions, and differing from those of the diseases of the spinal cord roots or of those of the primary trunks resulting through their junction.

From these plexuses spring longer and shorter branches, by which the nerve fibres, now finally divided, are led to their innervation areas, without undergoing any more anastomoses. These branches, in the narrower sense of the word, may be called peripheral nerves. The clinical disturbances which result from their lesion are again characterized by distinct groupings of their symptoms and by local diffusions. The diseases of these nerve trunks and of those of the first named plexus form the principal object in dealing with the diseases of the peripheral spinal nerves.

In the domain of the *cerebral nerves*, anastomotic formations do not play such an important part as they do with the spinal nerves. The trunks proceeding from the brain retain, for the most part, their essential composition until their peripheral distribution. Nevertheless, even some of these nerves possess anastomoses in certain places in their course, through which functionally important species of fibres leave or are joined to them. In this way particular varieties of clinical pictures are produced, depending on whether the nerve trunk be injured in one place or another, above or below an anastomosis. These symptom complexes afford us important aid for the finer diagnosis of the seat of the lesion.

A description of the *more intimate structure* of the peripheral nerves is unnecessary. It is assumed that the histology of the nervous fibres, their construction from axons, medullary sheath and neurilemma, is known. All fibres of peripheral nerves are constructed after one type. Only the olfactory nerve consists of non-medullated fibres. It is further assumed as known, that the nerve fibres are everywhere bound together to the nerve trunk or branch by connective tissue, which contains also the nutritive blood-vessels. According to experimental and pathological observations, the lymph spaces of the nerve trunk communicate with those of the central organ. It is a widely accepted belief that the connective-tissue capsule possesses its own sensory nerves, the so-called nervi nervorum.

Lately the question has been discussed, how the functionally different species of fibres are distributed in the peripheral nerves, whether they are all mingled together, or whether certain fibres subserving special functions, occupy special places in the nerve trunk. The adherents of this latter theory

have spoken of a systematization of the peripheral nerves. No conclusive results have been obtained so far and our answers to clinical pathological questions have not been enriched from this source.

What has preceded makes it clear that the diseases of the peripheral nerves have been defined as partial diseases of both peripheral neurons. From what follows, it will be seen that a rather different *definition* is somewhat more fitting, especially since the histological examination of some cases shows, that, indeed, not only certain parts of the neurons are affected, but that these in their total outspreading are anatomically changed. We therefore define the diseases of the peripheral nerves, as such diseases of the peripheral neurons as definitely start from certain primary points within the domain of the peripheral nerves.

Classification of Peripheral Nerve Diseases

We divide these diseases into two large groups: the destructive and the neuralgic diseases. The first group is marked by a decided underlying anatomical condition consisting of serious disturbances in the structure of the tissues and clinically through the preponderance of symptoms of lost function (Ausfallerscheinungen). In the neuralgic, on the other hand, we find no regular underlying anatomical condition, while peculiar pains characterize the clinical picture.

(I) THE DESTRUCTIVE DISEASES OF THE PERIPHERAL NERVES

A. General Part

(1) General Morbid Anatomy

The histological changes in destructive diseases of the peripheral nerve concern the nerve fibres and the nerve connective tissue. The *change of the nerve fibres* may be divided according to two primary types. One form may be designated as *Wallerian degeneration.* Since the investigations of Waller (1856) we know that after complete division of a nerve fibre, its peripheral part rapidly—in fact, within a very few days—undergoes a peculiar degenerative process. After some finer changes in the structure of the axon have taken place, the nerve fibre degenerates by a kind of segmentation into a number of fragments, which, at first larger, gradually diminish in size. An increase in the mass of their protoplasm and the number of their nuclei is produced in the cells of the myelin sheath. The fragments of the ruined fibres are gradually absorbed; nevertheless medullary detritus is found, even after the expiration of some months, in the myelin sheath (Fig. 26).

Investigation of the other important form of fibre disease begins with

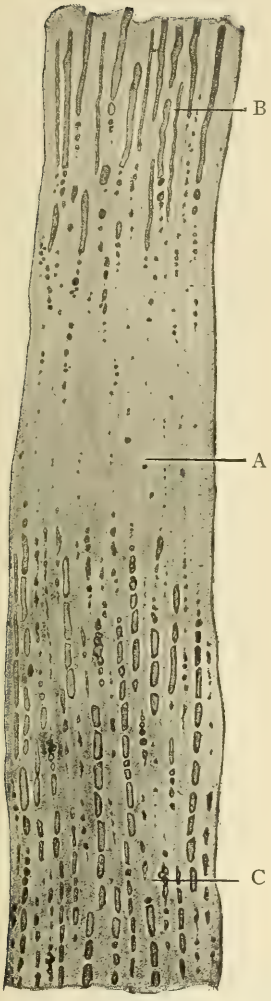


FIG. 26.—Wallerian degeneration. (*After Lugaro.*) Bundle of nerve fibres from the ischiadicus of a rabbit which was constricted four days. Osmium preparation. *A*, place of constriction; *B*, central section; *C*, peripheral section.

FIG. 27.—Degeneration of fibres of the type of periaxial medullary disintegration. (*After Stransky.*) Experimental neuritis through lead-poisoning. The disintegrated parts of the medullary sheath blackened by osmium.

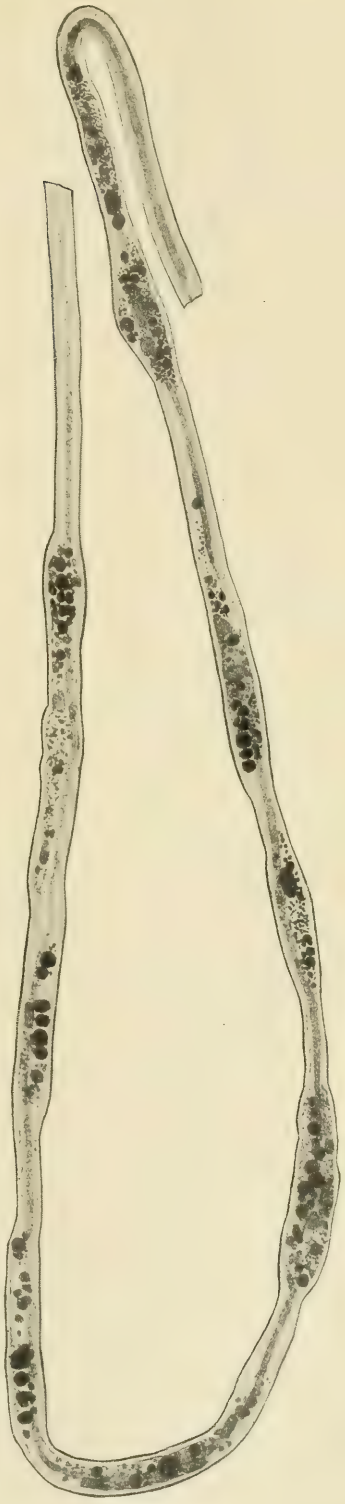


FIG. 27.

Gombault's work in 1880. We designate this form with reference to its most pronounced trait as the type of *periaxial medullary disintegration.* The normal medullary sheath of the axon shows, at regular short intervals, interruptions in its course. At these points the nerve fibres have waist-like constrictions, known as the nodes of Ranvier. Here the periaxial medullary disintegration frequently originates, first in the immediately adjacent sections; the outer layer of the neurilemma is disintegrated into more or less fine grains. This fine-grained disintegration then seizes upon larger sections of the fibres and pushes through the entire thickness of the medullary sheath. Here, too, by a process of proliferation, the cells of the neurilemma participate in the morbid process. Histologically, the axon undergoes changes only later. Whether the total destruction of the axon, and through this the local degeneration of the peripheral part and secondary Wallerian degeneration, is possible, is still under debate, but there is much in favor of this assumption. Repeatedly normal fibre sections between diseased sections have been observed in this form of diseased nerves (segmentary neuritis) (Fig. 27).

While Wallerian degeneration dooms the fibre to destruction, the changes in periaxial disintegration permit of a regeneration. In contradistinction to these two most important forms of fibre disease, the simple atrophying processes play only a subordinate part in the morbid anatomy of the clinical types of these diseases.

The *alterations of the nerve connective tissues* are either secondary or independent. In the first case, we are concerned with a mere increase, a sclerosis, such as always accompanies the degenerative diseases of the parenchymatous tissues. Less often we find primary interstitial disease, which usually affect the nervous elements also. Especially those pure inflammatory processes have to be considered here, which may arise in cases of hyperæmia, exudation, cellular infiltration and even with hemorrhagic and purulent changes.

In the most diverse diseases of the peripheral nerves we frequently find, moreover, peculiar *changes in the areas of the central sections of the affected neurons.* In the first place, in those diseases of the peripheral nerves which arise from toxic substances in the blood, pathological processes occur in the central organs, especially in the spinal cord, which, according to their kind and extent, must be regarded as independent complications. As a rule, no clinical interest attaches to them. Secondly, peculiar changes, which are evidently restricted to the central parts of the peripherally diseased neuron, may be observed in many cases. Above all, the root cells of the anterior horn and the spinal ganglia, are implicated in the disease. It has been ascertained through experimental investigations, that usually a histological change sets in after complete division of a nerve fibre in the appertaining ganglia cell, which consists chiefly in the destruction of the central Nissl

bodies, situated in the vicinity of the nucleus, and in an outspoken eccentric displacement of the nucleus. Analogous changes may frequently be noticed in clinical diseases of the peripheral nerves. While in most cases these adjust themselves again, very different, undoubtedly degenerative changes, and even total destruction of the cell may take place, if the nerve injury is very serious and permanent. Very instructive observations have been made on the spinal cords of persons who have suffered amputations. The cells of the anterior horn, corresponding to the amputated extremity, may atrophy or may remain intact for years. It is impossible to state with any degree of certainty, whether the one or the other will occur.

The proximal sections of the neurons, as well as the cells, may suffer trivial, or in case of the destruction of the cells, even considerable degenerative changes. Thus, for instance, dystrophic processes are observed in precisely those areas of the posterior tract of the spinal cord which are occupied by the central continuations of the peripherally diseased sensory neurons. Consequently the anatomic process in many cases presents itself as a general disease of the peripheral neurons. Merely histologically, it is then not always possible to recognize with certainty the primary attacking point of the injury. To decide in such cases whether a primary peripheral disease exists, it is necessary to refer to the clinical picture.

Even if the nerve fibres are completely destroyed, an anatomical and functional *regeneration* is possible, so long as the cellular centers of the affected neurons are preserved with the central region. Whether the new axon emanates from the proximal part and therefore, from the central cell, or whether it is autochthonously regenerated in the periphery, is at present the subject of an animated controversy. Perhaps the peripheral remnant of the destroyed nerve does more than merely serve as a guide for those fibres which newly originate from the center. At any rate the doctrine of the neurons, the theory of the unity of the ganglia cells and their continuations, can not be regarded as refuted in the chief points of its embryological and pathological foundation, and this theory alone is able, at present, to explain satisfactorily a great many incidents in the pathology of the diseases of the peripheral nerve.

(2) General Symptomatology and Pathological Physiology

The disturbances which are common to the diseases of the peripheral nerves, are determined by the nature of the fibres composing the nerve. Definitely known are motor, sensible and sensory, vasomotor and secretory fibres.

The most important symptom of a *lesion of motor fibres*, is the loss of voluntary motion, which may range through all stages from slight paresis to the total loss of motion, paralysis. Motor irritation phenomena are com-

paratively rare and have no special practical interest. Besides, it is exceedingly difficult to explain or to define them. A disease of the motor fibres should be assumed only if clinical symptoms of loss of function are present. In a few rare cases, even if the course of a single motor nerve, especially the ulnaris, is interrupted entirely, there need not be any paralysis in its innervation area, although as yet no experimental interpretation or satisfactory explanations have been offered for this occurrence.

Passive muscular contractures are observed to result from peripheral paralyses. In time the antagonists of the paralyzed muscles fall into a more or less extensive and permanent process of shortening and shrinking, which impedes also, to a corresponding degree, passive motion of the paralyzed group, and which may lead to considerable deformity. Another form of contracture, which affects the paralyzed muscles themselves, will be considered when discussing facial paralysis.

A detailed description of the disturbances of *electric irritability*, appearing in peripheral paralysis, can be found in this work under the heading "General Diagnostics." Only a few points, essential to an understanding of peripheral nerve diseases, are emphasized below.

In the lightest cases all disturbances of electric irritability may be lacking. Often there is a mere decrease—this happens more frequently than an increase for any length of time—in the electric irritability of the nerve-muscle area. In the majority of cases a reaction of degeneration (R. D.), in one of its numerous forms, may be shown. The sluggish, worm-like contraction of the muscle irritated by a direct galvanic current, is common to all these forms. These contractions, according to a generally accepted theory, are the deciding symptoms of R. D. It must be borne in mind, however, that sluggish contractions also occur in supranuclear paralyses (at least in a certain stage of the condition) occasionally in some of the affected muscles, and that the myotonic reaction may also produce deceptive sluggish contractions, which bear a striking resemblance to those of R. D.

About the relation of R. D. to peripheral paralysis, the following should be noted.

In some diseases, as, for instance, in lead poisoning, R. D. has been found even in muscles not affected by paralysis. This fact may occasionally acquire significance as an objective symptom of organic disease of the nerve-muscle apparatus (which otherwise could not be proven definitely) or, perhaps, as an early symptom of such disease.

As a rule, if paralysis is occasioned by lesion of the nerve, the point of lesion is rendered incapable both for the transmission of volitional impulses and of the electric stimulus. If the nerve trunk is excited electrically above the point of lesion, only those nerves can contract, whose motor fibres leave the nerve trunk between the point of lesion and the point of irritation. The nerve section located below the point of lesion, is at first both conductive and

irritable. After a short period of increase (which, however, can not always be noted), its excitability decreases rapidly. In the case of a complete division of the nerve it disappears entirely, approximately, on the twelfth day. The direct faradic irritability of the muscle is wont to decrease just as rapidly. It, too, disappears entirely in severe cases. The galvanic muscle irritability exhibits, in the majority of cases, an increase at the end of the second, or during the third, week, and this increase continues from three to nine weeks, after which it gives place to a decrease. The sluggish contractions occur during the period of increase as well as the period of decrease.

The conditions of the regenerative period are particularly interesting in those cases in which the irritability of the diseased nerve branch is entirely lost. Voluntary motion returns to the paralyzed area before its excitability to an electric current. Evidently the nerve becomes able to conduct volitional impulses before it is able to respond to an electric stimulus. In proper cases, observations may be made which show that conductivity for an electric stimulus returns before electric irritability. It is found, for instance, that when a nerve has been completely divided, an electric stimulus applied at the time of the return of active motility above the point of lesion, causes the muscles to contract, while a stimulation below that point is without effect. The R. D. outlasts the paralysis for a considerable time.

If it were possible to place the various symptoms of the disturbances of electric irritability, especially of R. D., in relation to certain physical or chemical changes in the nerves and muscles, it would mean a great step forward towards the establishment of a satisfactory theory for these symptoms. Of course, such knowledge would be of inestimable benefit for establishing a diagnosis, and, above all, a prognosis. Concerning the changes which affect the nerve, it may be said with a tolerable degree of certainty, that the irritability depends on the integrity of the medullary sheath, while the conductivity is dependent only upon the axon. The facts adduced above are in complete accordance with this view. The conditions during regeneration especially are thereby made comprehensible.

If a nerve trunk is completely divided, the axon is the first part of the destroyed section to regenerate, by, it may be assumed, an outgrowth from the central stump. The new growth of the axon is accompanied by the return of conductivity for volitional impulses, which was discussed above, and of electric irritability. It is only after a long time, however, that the newly formed nerve is clothed with a sufficient medullary sheath, and it is in accordance with this that the response of the regenerated section to electric irritability, its electric stimulation, returns much later than its conductivity.

It must be assumed, that the altered irritability of the muscular substance also depends on variations in its more intimate structure, although no generally recognized opinions concerning the nature of these variations exist.

It is certain, however, that the older theory, according to which the coarse degenerative changes correspond to R. D., is untenable.

The muscle that responds with R. D. to an electric stimulus, responds frequently to a mechanical stimulus—such as a slight tap with a percussion hammer—with a sluggish contraction, the so-called *mechanical* R. D. In this case, too, quantitative changes of irritability are found.

In a few rare cases, it has been noted that the paralysis continues, notwithstanding the return of electric irritability. In many such cases it must be assumed that the cause for the continuing deficiency in voluntary motion is not to be sought in the area of the peripheral neuron. If, despite the fact that the neuro-muscular apparatus is evidently restored, the patient does not regain control over the muscles concerned, it is possible that the cause may be found in the domain of psychology. *Oppenheim* has spoken of a habit paralysis. In other cases this symptom is to be explained differently. (See section dealing with the course of facial paralysis.)

The symptoms of loss of function in the *sensory area* are decidedly less important than those in the motor area. When a mixed nerve becomes diseased, the motor manifestations of lost function appear in the foreground, while the sensory occur later and are less pronounced. They are restricted to parts of the innervation area, much more frequently than the disturbances of motility (these partial disturbances of motility will be discussed later in the chapter on diagnosis). Occasionally they are missing entirely, but this is very rare; especially rare during an entire course of a serious disease. Sensory function frequently returns long before the motor.

Numerous attempts have been made to explain these peculiar facts, but none of them is quite satisfactory. The theory of the less liability to lesion of the sensory fibres is really no more than begging the question. It may be taken for granted that the double support rendered the same sensory area by different nerves, and anastomotic formations between branches of different nerves, play a very important part, varying in different individuals.

The different cutaneous sensation qualities, such as the sensory perceptions of touch, pain and temperature, are, in most cases, equally impaired; but exceptions to this rule may occur, in which cases the perceptions of pain or temperature may suffer chiefly or exclusively.

Slight as is today the basis on which to found an explanation of these occurrences, it is nevertheless safe to say, that they appear not alone in diseases of the spinal cord, but also, though much more rarely, in peripheral affections.

Comparatively frequently, besides a considerable lowering or disappearance of tactile and thermic excitability, *hyperalgesia* is found. A slight pricking with a needle, light pressing or stroking of the skin, or stimulation with feeble electric currents, even the pressure of the clothes, gives rise to violent pains, while mere contacts are not perceived. Spontaneous pains

may also occur in these cases. This condition has been designated as anæsthesia dolorosa. It is found often in diseases resulting from alcoholic or arsenical intoxication and in herpes zoster, more rarely after injuries of a nerve.

Retardation in *perception of pain*, which is well known in tabes, also occurs in peripheral diseases. A definite explanation of this phenomenon is wanting in both cases.

We have just touched upon the *irritation symptoms*. They play a rôle of far greater importance in the sensory than in the motor areas. An actual hyperæsthesia, it is true, has not been noted with certainty aside from the previously mentioned hyperalgesia. On the other hand, paræsthesia and pains play a rather important part. The first manifests itself as formication, pricklings, itchings, chilliness, and other similar unpleasant sensations. The pains are felt partly in the course of the nerve, partly in the area it supplies. Besides spontaneous pains, tenderness to pressure in the nerve trunks plays an important, though formerly greatly overestimated, part. The pains, which during disease of the nerve trunk are projected into the area of peripheral distribution, may be traced to irritation of the long sensory fibres which course in the nerve. The nervi nervorum are probably largely responsible for the production of the other symptoms spoken of. We know scarcely anything concerning the disturbances in sensation of the deeper parts (ability to distinguish differences of pressure; susceptibility to passive joint-movement) in peripheral diseases. Especially little is known about the deeper innervation areas of the single nerves.

There occur cases of disease of mixed nerves, in which, contrary to the above stated rule, the sensory loss of function is greater than the motor loss. Nothing definite is known about the conditions under which this occurs. Sometimes the disturbances in motility show the character of those present in locomotor ataxia, which according to the theory established by Leyden is caused by the loss of centripetal impulses (neurotabes peripherica).

Since in disease of the peripheral nerves the motor as well as the sensory limb of the reflex arc of the tendinous, periosteal, and cutaneous reflexes are affected, it is self-evident that disorders of the reflexes (decrease and loss) will occur. In general, one may say that the reflexes cease very early. The loss of reflexes may often be the first objective indication of a destructive nerve disease, just as it commonly outlasts all the other disturbances, and may remain as a last and frequently permanent residuum. Cases occur, on the other hand, in which the reflex, in spite of an indubitable disease of the area of its reflex arc, persists remarkably long. Aside from its occurrence as a transient initial symptom in some cases, a reflex increase is an exceedingly rare occurrence, and is always a strong argument against a peripheral nervous disease. Of course only those areas are meant which are actually affected by the disease. An increase, for example, of the Achilles tendon or

patellar tendon reflex in a pure peroneal paralysis is a common occurrence, as the peroneus nerve is not at all essential for the formation of these reflexes.

In a number of infectious fevers it frequently happens that during the febrile stage, the tendon reflexes of the limbs disappear, at least temporarily. This can be observed in lobar pneumonia, in typhoid, in diphtheria, and even in ephemeral fever. The same thing occurs also in a number of toxic conditions (alcoholism, diabetes mellitus), though in such cases the loss frequently is permanent. It is not yet known whether in these cases the anatomic lesion is to be sought in the area of the peripheral nerves.

We know little definite about the *vaso-motor disturbances*. It is assumed that the peripheral nerves carry both vaso-constrictor and vaso-dilator fibres, thereby making it possible to understand many clinical symptoms which accompany peripheral nervous diseases. Here and there redness and heat are observed, in other cases the diseased parts are cold, pale and cyanosed, which can, in part, be definitely ascribed to the disturbances of the innervation of nerves. No considerable practical interest attaches to these disturbances.

Secretory disturbances will be treated minutely in the discussion of the paralysis of the seventh and ninth cerebral nerves. In passing, only a word regarding the production of sweat. With the motor peripheral nerves course the secretory nerves for the sweat glands. Destruction of the peripheral nerves may lead to the complete loss of sweat secretion. In the early stage of peripheral diseases and in partial lesions hyperhidrosis, which may be classed as an irritation symptom, sometimes occurs.

Of the **trophic disturbances** those of the musculature are by far the most important. Atrophy of the muscle in diseases having an acute onset develops some time after the paralysis, while in diseases with a more chronic course, it may develop at the same time as the paralysis. As stated above, definite histological characteristics of atrophy dependent upon disease of the peripheral nerves are up to the present, unknown. The usual microscopic methods merely show the ordinary characteristics of "simple atrophy." It may be assumed as likely, that the trophic influence of the peripheral motor neurons coincides exactly with its functional significance. All irritations which are transmitted to the muscle through a nerve tract, the psychomotor and reflex impulses, ultimately take their course over the cell of the anterior horn and the peripheral motor nerves. If this course is interrupted entirely, the muscle is cut off from all nervous irritation, while in supra nuclear paralysis or interruptions of the pyramidal tract, irritations are still transmitted to the peripheral nerves through other supervening neurons. In this way the ceteris paribus, very severe, even complete, atrophy of the muscles in many peripheral paralyses is explained, in contradistinction to the usually far less pronounced atrophy in supra nuclear paralysis.

As to the contractures of the musculature some preliminary remarks have already been made.

Of further trophic disturbances, those of the skin are particularly noteworthy. Not infrequently the skin becomes peculiarly glossy, thin and smooth, while the nails, on the other hand, lose their lustre, become cracked, of irregular growth and sometimes crooked, so that a kind of onychogryphosis results. Occasionally inclination to serious cutaneous diseases, eruptions, formation of ulcers and anomalies in hairgrowth are observed. For an explanation of edema, which is sometimes observed, it is necessary to take into consideration the immobility of the parts concerned.

As a peculiar cutaneous affection depending upon an affection of the peripheral sensory neurons, herpes zoster is to be mentioned. It is chiefly found accompanied by neuralgia and will be described briefly with it.

If the peripheral nerves of an entire extremity suffer a serious and extended lesion in earlier years, the affected limb is retarded in growth in all its parts. But even in similar diseases of adults, acquired later, a slight degree of bone atrophy will often be revealed by the X rays. The joints suffer especially under the influence of immobility.

Besides the above there are a number of trophic disturbances which have been designated as casuistic rarities. They are numerous and heterogeneous but no practical interest is attached to them. In general, great deviations from the above types are very uncommon.

The essentials concerning the neuro-physiological origin of muscle atrophy have been stated above. Our opinion concerning the manner in which the other trophic disturbances originate, is still more pronouncedly provisional. At all events, nothing forces us to the conclusion that there are, besides the other fibres, also trophic fibres in peripheral nerves whose only task would consist in influencing the nutritive condition of the tissues. It is much more likely that these disturbances are explained part by the injurious effect of the inactivity of the parts, more precisely by the loss of centrifugal impulses, partly by the impairment of certain regulatory antagonists in regard to external injuries on which the integrity of centripetal tracts depends. In the first place the cessation of sensation must be considered. In fact many trophic disturbances appear especially in those diseases resulting in grave losses of sensory functions. True trophic changes in joints occur only in diseases in which there are sensory disorders. Muscle atrophy occurs also where sensibility is normal.

(3) General Diagnosis and Prognosis

Destructive processes of the peripheral nerves can, as a rule, be diagnosed only in those cases in which clinical symptoms of lost functions exist. Where no paresis or paralysis, no hypæsthesia or anæsthesia exists, it will

be very difficult to reach even a partly certain diagnosis. One should be especially cautious in not overrating the diagnostic significance of pains and tenderness of the nerve trunks on pressure. The latter is an equivocal sign and is found under manifold circumstances. In many nervous persons all the nerve trunks react to pressure with pain sensations. In various affections of the joints the same symptom is found, more or less pronounced. Very rarely pains appear in such characteristic ways and places as to deserve any diagnestic consideration. The typically neuralgic pain, which will be considered in another chapter, does, indeed, unquestionably indicate a disease of the peripheral nerves, but points to so-called neuralgic changes, not to destructive affections. The relation between neuralgia and destructive nerve diseases will be treated in the chapter on neuralgia. In most cases the existing pains are not very characteristic.

The correct diagnosis depends on the accurate interpretation of the symptoms of loss of motor and sensory power.

In the first place, paralyses possess certain common traits, peculiar to the diseases of the entire peripheral neurons, and secondly they are characterized by certain extensions of the disturbances which correspond to the innervation area of the peripheral nerves. Taking these facts as a criterion will in many cases facilitate the diagnosis considerably.

The following may be mentioned as general neuron symptoms. The peripheral paralysis is in so far absolute as the muscles not only fail for certain functions, as in many supra nuclear paralyses, but are also in the same degree cut off from all nervous stimulating impulses. Consequently the so-called associated movements, which are frequently seen in central paralyses, are lacking. A peripherally crippled hand will not reveal a contracting movement, even after strong exertion of the healthy hand.

In certain cases, a sort of associated contraction sets in sometimes in a peripherally crippled muscle, if intact nerves in its immediate vicinity are innervated. This constitutes a rare, almost indefinable symptom, which has been noted in the area of the ophthalmic muscles. Another very singular form of associated contraction in paretic and paralyzed muscles will be spoken of in the chapter on the course of facial paralysis. These facts, however, do not destroy the value of the above cited rule.

Peripheral paralysis is flaccid. In parts not wholly paralyzed, a certain amount of resistance is sometimes noted in passive motions, if these awaken pain, though a real spasm without impairment of reflexes is never produced. The tendinous, periosteal and cutaneous reflexes depending on the diseased nerves are weakened or abolished. The genuine Babinski reflex does not occur. (Concerning a pseudo-Babinski the reader is referred to tibial paralysis and sciatica.) The muscles, at least in the serious cases, become very atrophic. Electrical examination as a rule reveals R. D. But not in all cases will the above mentioned criteria permit of a neuron diagnosis

with certainty. In the chapters of this book dealing with the subject, it will be shown that central paralysis may also be flaccid, and be accompanied by reflex losses, and that it, too, may cause muscle atrophy to a great extent. On the other hand, the muscle atrophy in peripheral paralysis may be remarkably slight, as it is generally in the lighter cases. But even in severe peripheral paralysis, in multiple neuritis for instance, the muscles are frequently, from the clinical standpoint, not at all atrophic, sometimes even hypertrophic. In a few cases it was possible to trace the cause of this condition anatomically to a lipomatosis of the musculature.

In contradistinction to these difficulties, the diagnostic significance of the extension of the disturbances to the motor and sensory innervation areas of certain peripheral nerves, in the more restricted sense, or of certain branches of the nerve plexus, by which for one familiar with these areas the diagnosis at once suggests itself, is all the more important. More about this may be found in the description of the various paralyses.

A few special difficulties which may obstruct a proper diagnosis, and are particularly liable to mislead a novice, should be indicated here. We shall not attempt, however, a discussion of the rather rare complication of central by peripheral disturbances.

If very many peripheral nerves are simultaneously affected, the symptoms of lost function may be so extensive as to preclude the possibility of recognizing a typical distribution by peripheral areas. If entire extremities, even all extremities of the body are affected, it is yet possible, that by reason of the disproportionate affection of the various peripheral nerves the peripheral distribution type may be clearly marked in a few places, although this sometimes is not the case. Then the symptoms we have described as neuron symptoms, are of predominant importance in determining the diagnosis. For the diagnosis of a peripheral disease, in contradistinction to a spinal affection of the same neuron, besides the certain features of its entire clinical picture, ætiological factors, condition of its development and course, which are characteristic of certain types of diseases, the following is, above all, important. Almost invariably motor and sensory disturbances co-exist in peripheral diseases, just as in the most important form of disseminated spinal affections of the peripheral motor neuron, sensory disturbances are, almost without exception, lacking. In other spinal diseases, besides symptoms of the peripheral neuron disease, there appear typical, specifically central or spinal disturbances, foreign to peripheral diseases; symptoms of a lesion of the pyramidal tract, bladder disturbances, etc., which unite with those into characteristic clinical pictures of the destruction of certain spinal areas. (See differential diagnosis of multiple neuritis.)

But even in slightly disseminated processes difficulties may arise, since in diseases of the peripheral nerves the symptoms of lost functions may be confined to certain districts of the innervation area. First the paralysis may

exhibit itself in different branches of a nerve consecutively, or it may continue to be restricted to single branches. The preference which the disease manifests for certain parts of the nerve area, is in many cases dependent not on anatomic, but on functional conditions, so that under the influence of any injuries, those parts will be affected primarily or exclusively, from which especially strenuous and continued service is exacted. Furthermore, one sometimes sees a predilective or exclusive disturbance in the remotest part of the motor and sensory innervation area of a nerve, while the nearer parts remain intact. This is said to occur also in cases where the lesion has doubtless affected the main trunk. It has already been noted that the sensory disturbances are frequently, almost regularly, less pronounced than the motor. It is very seldom that sensory disturbances are lacking completely, though it, too, occurs, and mostly in light cases. Many diseases of the nerve trunks exhibit only functional disturbances of certain fibres functionally belonging together and terminating in the nerve, whereby a "systematizing tendency" is revealed. A classical example is afforded by many cases of oculomotor trunk affections, in which only the external, not the internal, eyemuscles are paralyzed. The explanation of such occurrences is still a matter of controversy. After what has been said, it is obvious that it is not always possible to localize definitely intra vitam a disease of the peripheral nerves. However, those are comparatively rare exceptions.

The prognosis of a peripheral nerve disease must be viewed from two standpoints. First with relation to existing causative factors. If one is certain that the injurious influences are at an end, that no new noxious factors of any kind can influence the diseased parts, it is possible to make a prognosis, not conclusively, it is true, but with a reasonable amount of certainty, according to the degree of the existing disturbances. A test of electric irritability is of great value in such cases. Though one may frequently hit upon a correct diagnosis without such a test yet, upon the whole, it will be practically impossible to make a fairly definite prognosis without careful electro-diagnosis. As stated above, an electric test reveals the extent of anatomic destruction. It must be borne in mind, however, that usually the disturbances of electric irritability are only sufficiently developed to permit of definite judgment, two, or even three, weeks after the injury has begun its work. Slight paralyses without any pronounced disturbances of the electric irritability may recover in a few weeks or even days. If R. D. be present, however, it will always require many weeks, sometimes even several months, for complete restoration. The more the electric irritability of the nerve trunk has decreased, the less favorable the prognosis becomes. When the nerve is mechanically divided, the prospect of reunion between the diseased and the peripheral part affects the prognosis considerably. Concerning its importance more will be said in another chapter.

(4) Classification of the Destructive Diseases

The destructive diseases may be divided into two large groups. One group comprises those cases, in which single peripheral nerves are injured, and those rarer cases of multiple disease, in which the multiplicity bears the character of being more or less accidental, so that no definite system of the extension of the disturbances may be recognized. In a great majority of these cases a definite local cause of the disease may be proven; in the remaining cases, such a local cause is to be assumed, if the existing localization is taken into consideration.

In the second of the main groups an absolutely certain, orderly, more or less strictly bilateral, symmetrical extension of the disturbances occurs and, as a rule, the resulting typical course of the disease is a very pronounced feature. These clinical pictures are the result of internal injurious influences of a general character which reach the nerves through the bodily fluids.

It has been repeatedly attempted to make, besides this classification, still another main division, namely, according to pathologic anatomic viewpoints, so as to separate under the name "neuritis" the pure inflammatory processes from the rest. This attempt has proven a failure, and precisely there, where we possess accurate pathologic anatomic knowledge, which is not always the case in our field. We shall not here point out the difficulty of defining the "pure inflammatory," but the following must be emphasized.

In the second main group of our division, the typical and regular alteration in the nerves, bears, as we now definitely know, a purely degenerative, but certainly not an inflammatory, character. Yet we find in some cases which neither clinically nor ætiologically are to be distinguished in any way from the others, besides the usual processes, purely inflammatory foci in several places. But surely no subdivision ought to be based upon this.

From the standpoint of nomenclature, therefore, there will scarcely be any objection to the retention of the term "polyneuritis" to denote all cases in this group, and from the standpoint of language, too, the word is absolutely correct, if one recalls the purely adjectival meaning of the word neuritis. As one no longer thinks only of purely inflammatory processes when hearing the word "nephritis," so one will also have to deprive the word "neuritis" of the inflammatory characteristics, which have frequently been assigned to it.

Not many attempts have been made to subdivide our first group since it was realized that such attempts would be impracticable from the viewpoint of morbid anatomy and would show all the deficiencies of an artificial system, though it has been divided in such a way as to separate diseases resulting from acute mechanical trauma as a specific group from the others classified as "neuritic" diseases. But even in this, some artificiality may be

clearly detected, as will be evident from the following description, which disregards such division entirely.

In the first place, therefore, we shall deal with the diseases affecting single peripheral nerves, peripheral paralysis in the more restricted sense, mononeuritis in the broadest sense of the word, including also mononeuritis multiplex, and in the second place we shall deal with polyneuritis.

B. Special Part

1. Diseases of Single Peripheral Nerves

General Ætiology

Of greatest ætiological interest are the *local injurious influences.* The relations to mechanical trauma are remarkably diversified. Mechanical injuries act directly or indirectly on the nerves. The direct traumatic effects are of the most varied degrees from a comparatively light pressure to complete division, tearing asunder, cutting or crushing of the nerve. The indirect traumatic effects are produced by such causes, for instance, as an excessive motion of the body, producing an undue stretching of the nerve. This will be gone into more minutely when discussing paralysis of single nerves.

Indirectly, trauma may be responsible for peripheral paralysis in three ways. In the first place, long continued mechanical irritation, such as the pressure of crutches, may in time lead to degenerative processes and nerve paralysis (so-called traumatic neuritis). But even a single mechanical insult seems to be able to give rise to similar diseases. The trauma is at once followed by pain. Only after some days—or weeks—the symptom of lost function develops. Secondly, conditions resulting from an originally non-nervous lesion may cause mechanical nerve injuries. After fracture of a bone it is sometimes observed that nerve trunks not primarily affected may be injured secondarily by bone fragments. Scar tissue or the outgrowth of callus may lead to direct-pressure injuries by growing around and constricting a nerve, or nerve trunks may be forced into such unfavorable positions that they are exposed to injuries whenever a member is moved. Nerve dislocations, by which the nerve is displaced from its normal position, through traumatic influences, act in the same way. Under such conditions, an old trauma may lead to paralysis even after several years (late traumatic lesions). In the third place, finally, so-called ascending neuritis must be considered. As a result of small infected wounds, splinter injuries, etc., especially of the hand, grave neuritis of the nerve, in whose innervation area the wound was located, develops in rare cases.

Other affections in addition to trauma may cause mechanical injuries to nerves. Tumors, in the broadest sense of the word, and bone disease, may all lead to paralysis of neighboring nerves through pressure.

Malignant tumors may also affect the nerve directly; inflammation in its proximity may spread to the nerve itself and either cause an inflammation, or its degeneration through toxic influences. Ulcers and other cutaneous diseases lead now and then to degeneration of the sensory terminal fibres in their own area and that in their immediate vicinity—a most interesting fact theoretically, but one which has been but little studied.

In order to explain a few peculiarities of the clinical picture, a separate short discussion will be devoted to tumors arising from the different anatomical elements of the nerve trunk itself.

A peripheral paralysis occasioned by local chemical injury is sometimes seen after a bungling injection of ether or camphorated oil containing ether, made in the immediate vicinity of a nerve trunk. Some observations would seem to make it probable that absorption by the skin of bisulphide of carbon —an injury to which workers in rubber are often exposed—may cause local neuritis.

Local thermic injuries play only a subordinate part. When discussing facial paralysis we shall meet with conditions which may be regarded as arising from local colds.

It is said that arterio-sclerosis of the vasa nervorum may lead to degeneration of the nerves.

As a final local cause overexertion of certain parts of the body in particular occupations must be considered. It will be necessary, however, to discuss briefly the peculiar so-called occupation paralyses in a separate paragraph.

Besides the local influences, *general* injuries acquire some significance, even in the ætiology of the diseases of single peripheral nerves, though it can not be said they they play more than a predisposing rôle. The same things will have to be named here as are enumerated in the ætiology of polyneuritis: infections and toxic influences, disorders of metabolism, pregnancy and the puerperium, and debilitated conditions. Persons affected with tabes reveal a peculiar inclination to peripheral paralysis. In all such cases it is advisable, however, to assume a locally acting disturbance besides the general predisposing causes. If, in a few cases, it is not possible to prove it, it should be remembered that with a strong predisposition under certain conditions, trivial local influences which may be easily overlooked suffice to excite disease. Sometimes, at least, conjectures may be formed concerning their nature. If, for example, an alcoholic after the passing of the delirium has a radial paralysis, it can scarcely be amiss to assume a traumatic cause in addition to the toxic predisposition.

Pathological Anatomy

Simple disintegrative processes of the nerves are much more frequent than purely inflammatory changes. The latter are found especially in those

cases in which the vicinal inflammation has extended to the nerve (extended lymphogenous neuritis), in neuritis emanating from infected wounds and in local influences of chemical poisons. The nerve in such a case is turgid, reddish, while the microscope reveals hyperæmic and exudative changes, and cellular, even suppurative infiltrations. Soon also, naturally, the parenchyma participates in this process. Occasionally bacteria may be found in the nerve trunk. To distinguish between perineuritic, interstitial neuritic, or nodular forms according to the degree to which the perineurium or the endoneurium participates, or according to the form of the nerve swelling, would be useless.

The parenchymatous degeneration which, as was said, in most cases exists without actual inflammatory complications, bears in most of the lighter cases the character of periaxial decay. This has been shown to be true especially of the lighter traumatic influences. Though the axon is surely not anatomically destroyed, its conductivity for the will impulses, at least, may be affected temporarily. In such cases the mobility returns very soon after the causative disturbance has been removed. Even if the duration of the injury be longer (as in pressure due to callus and the like), the disease may remain in this lighter stage, and under proper treatment permit of a very favorable prognosis.

If the local injury is greater, and the axon is entirely destroyed, the peripheral nerve section is doomed to Wallerian degeneration. The regeneration will take the longer the more centrally the interruption of the nerve has taken place. The reunion of the central and peripheral parts, even if produced only by interposition of granular tissue which paves the way for the newly formed fibres to the peripheral part, is an important factor in the process of regeneration. Should the reunion not take place, the regenerative efforts of the central end will lead to the formation of what is known as amputation neuromata; knoblike swellings that form clinically most irritating structures, which may cause violent pains.

Those very rare cases in which functional restoration has been reported without the reunion of the two nerve ends are perhaps best classed with those equally rare cases, already mentioned, of the complete division of large nerve trunks without clinical symptoms of lost function.

The *clinical picture* will be considered here only very briefly. Those symptoms which occur in common in destructive diseases of the peripheral nerves have already been discussed in the general part and the clinical picture in diseases of different single nerves will be shown in the next section. It may suffice to state summarily once more here, that in paralyses of mixed nerves, the characteristically grouped motor symptoms of lost function together with the accompanying disturbances of the reflex function and of the electric irritability, stand almost without exception in the foreground. As a rule, the sensory symptoms of lost function, according to the

principle cited before, yield to the motor ones; still they belong to the cardinal symptoms, even though they must often be sought for. More often symptoms of sensory irritation are lacking. No general rule can be laid down regarding their occurrence or non-occurrence. It is to be remarked, however, that their absence is the rarer, the greater and more continuous the irritation of the nerve trunk dependent upon anatomic processes is. It would be presumptuous, nevertheless, to draw from the sensory irritation symptoms definite conclusions concerning the finer anatomic nature of the nerve disease. Least of all is it possible to establish a rule about the pressure sensitiveness, which sometimes extends to the musculature of the diseased region. In the pure inflammatory processes, cord-like or knotty constrictions of the nerve trunk may sometimes be felt. Whenever fever occurs it indicates that an infectious process is producing the paralysis.

For a discussion of the *diagnosis* of the paralyses of single nerves, which is usually very easy, the reader is referred to the general part, and for some specific peculiarities, to the following section. Only a few remarks regarding differential diagnosis are made here. We shall only mention here the great demand that complications blurring the clinical picture of the peripheral paralyses, because of central organic and hysteric disturbances, make on diagnostic acumen.

Real difficulties arise now and then, especially in lighter cases, when the symptoms of the paralysis are not very grave, its boundary not clearly defined and when it affects only a few of the muscles subserved by one peripheral nerve. Differentiation of spinal affections of the peripheral motor neuron may then be rather difficult, and all the more so, because the innervation areas of some of the spinal segments are very similar to those of some of the peripheral nerves. Even when the disturbances of sensation do not correspond to a whole peripheral nerve area, but are confined, say, to a distal part of one, it is not always possible to base a decision on this, as in spinal diseases also the disturbances of sensation may be restricted to distal parts of the innervation area. In cases in which the knowledge of certain ætiological factors, or the course of the disease, or the finding of decisive symptoms in other areas, does not enable a sure diagnosis, it will have to remain in abeyance.

Experience shows that certain non-neurogenic contractures, atrophic and paralytic conditions, sometimes lead to an incorrect diagnosis, which, however, might almost always be avoided in these cases. Tendon and fascia contractions, joint anomalies and cicatricial processes, may produce deformations of the limbs, which superficially resemble the contracture produced by peripheral paralyses. A careful examination will enable one to decide whether the situation of the contracture is conditioned upon a loss of function in a definite nerve area. Of course, extensive fixation of the part may make the investigation more difficult, especially in inveterate cases, and

it will then become necessary, besides considering the contractibility of the muscles, the reflexes, the electric reaction and the sensibility, to consider whether the existing deformities may at all be traced, according to their form, to peripheral paralysis (for the form of contractures in peripheral paralysis is usually very characteristic), and whether other causes for the deformity are to be found.

It is usually very easy to determine correctly arthrogenous or arthritic atrophy, because of its characteristic distribution in certain definite muscles in the vicinity of a diseased joint. Usually the extensors of the joint are affected. Atrophy of the quadriceps femoris in diseases of the knee and the predominant atrophy of the deltoid and supra- and infraspinatus in arthritis of the shoulder joint is especially important. The functional impairment may attain to actual paralysis. The tendon and periosteal reflexes are in such cases always exaggerated.

Ischæmic paralyses and contractures result when the blood supply to a certain muscle area is seriously obstructed by too tight bandages or, as occurs more rarely, through lesion of blood-vessels. If relief is not had soon, the muscles fall into a paralytic and degenerative condition resulting in a rigid contracture, which is not dependent upon nervous lesions. It has been especially noted in the upper extremities and is minutely described in surgical text-books.

These and other myopathic paralyses and contractures are distinguished from similar peripheral neuritic affections, not only by the ætiological peculiarities, but also by the absence of pronounced sensory symptoms of lost function, by the regular absence of R. D., and by the peculiar distribution, which is not confined to the innervation area of peripheral nerves.

If careful consideration be given to the nature of the symptoms of lost function, and if further, the anatomic extent of the symptoms which are dependent upon the extent of the innervation areas be as carefully determined, the diagnosis and differential diagnosis will fail one only in very rare cases.

Prognosis.—What was stated above concerning the different ætiological factors will serve as an ampler illustration of the fundamental principle already referred to in the general part, namely, that the prognosis is affected decisively by the nature of the primary injuries.

A few viewpoints not here discussed, will be dealt with in the following therapeutic section.

General Therapeutics

The *prevention of peripheral paralysis* not infrequently occupies the attention of the physician. Both the surgeon and the obstetrician very frequently are compelled to endanger the peripheral nerves in various ways. Even if it is now and then, unavoidable to injure a nerve for the sake of

attaining more important ends, it might more frequently be avoided, if only the danger were considered. In making hypodermic injections, one ought not to select a place near a large nerve-trunk. In prescribing medicines, the possibility of producing hazardous conditions (as from the too long continued use of arsenic) must be considered. Of great import is the prophylaxis, depending upon the hygiene of the trades or occupations in which poisons are used. As to predispositions it is necessary to remember the special liability to lesion of the nerves of these individuals with regard to local influences (pressure paralysis in cases of tabes and in alcoholics). These suggestions will suffice here. The preceding ætiological section furnishes some further points, and in the following chapter special cases will be mentioned, in which the especial attention of the physician is essential.

The **actual treatment** begins with an endeavor to remove the original noxious factors as far as possible, or to prevent their further spread. Further procedure depends on the nature of the injury. If a nerve is mechanically divided, surgical aid, if at all possible, should be rendered at once. In other cases, one should, in the early stages, confine one's self to preserving in its broadest meaning the diseased part, avoiding all injurious influences, and to symptomatic alleviation of the most severe troubles. Later, one's efforts may be more directly aimed at the preservation and restoration of function. Generally speaking, it may be said that this more active method of procedure is in order only when the regressive changes will positively not progress any further. In paralyses caused by a single acute insult this method may be started about the third week.

General Measures.—In the early stage the diseased part should be kept absolutely quiet, be put in slings, splints, bandaged in cotton, and preserved from shocks (especially of the nerve trunks) by proper padding. The warmth generated thereby, is usually most beneficial.

Anæsthetic areas should be particularly guarded. Mechanical and thermic injuries, which the patient does not notice because of the anæsthesia, may lead to serious injuries, difficult to heal (burns caused by hot-water bottles).

That the general health must be carefully observed and taken care of, is not at all merely a schematic phrase, but in our field, particularly, is of the greatest importance. Experience has shown that the prospects for the regeneration of destroyed nerves depend considerably on the general condition of the patient. Because of this, general medical principles have to be followed. Anæmia, disturbances in metabolism, enfeebling diseases of any kind, should be treated carefully; the expenditure of strength, as far as possible, reduced, and the diet regulated.

Toxic injuries, whether occasioned by one's trade or occupation or by one's manner of living, should be strictly avoided, even if they were not a factor in producing the disease. Patients who imagine that they may in-

dulge their taste for alcohol during the treatment, have little prospect of recovery, and had better be discharged from treatment.

Medicines are to be considered first for the causal treatment. It is usual to point out, that in combating infectious diseases and conditions due to intoxicative processes, it is only necessary to proceed with suitable remedies, such as iodides and mercury, for syphilis; quinine for malaria; iodide of potassium for lead poisoning. But it is very seldom that these factors enter into the question at all—more frequently they lead, not to paralysis of single nerves, but to polyneuritis. Much more important with us is the use of medicines with regard to the individual symptoms. To reduce the pains we give the numerous well-known antipyretic and antineuralgic agents, concerning the prescription of which the reader is referred to the neuralgias. Morphine should be used very seldom.

Besides various other reasons against the use of vesicants and derivants in painful conditions, it may be urged that the injured skin may afterwards interfere with the electric treatment. If it is desired to administer a mild cutaneous irritant beneath the bandage, mistura oleoso-balsamica or a 10% menthol oil should be applied to the aching parts. If an intense inflammatory swelling occurs near a nerve trunk, an attempt at relief may be made by applying a few leeches. After the acute stage has ended, alcoholic and other embrocations may often be recommended in combating the paræsthesia. (Spirit. camphorat., 5% menthol in alcohol et al.)

Of those remedies to which a tonic action upon the nervous system is ascribed and from which for that reason a direct action upon the regenerative processes is to be expected, preparations of strychnine may be tried first. They are best given in the shape of so-called tonic pills (Erb).

℞. Ferr. lactic.,
Extr. chin. spirit. . . . . āā 6.0 (gr. 90)
Extr. nucis vom. . . . . 0.6 (gr. 9)
F. pill. LX S. 3 times daily a pill.

Or strychnin. nitrat. may be given by hypodermic injection, at first three times a week, after that once daily 1 mg. The dose may gradually be somewhat increased.

Hydro- and thermotherapy have an extensive field of usefulness. In the acute stage Priessnitz's compresses, which will usually have to be applied warm, may be tried alternating with dry dressings. Other forms of local devices which retain the heat may be tried. Poultices, cataplasms, thermophores will very often have a favorable action upon the pains. Cold applications are not so often beneficial. Should this be the case, water or ice bags, ice cataplasms and especially compress bandages may be applied, in which the limb is bandaged with a moist linen bandage, which is kept wet by frequent moistenings. Protracted cold applications are in general to be re-

garded merely as a transiently admissible symptomatic treatment in the acute stage.

Baths of the entire body or of the diseased part may be used advantageously in any stage of the disease. Baths of so-called "indifferent temperature" (96° F., 35° C.) have a quieting effect upon sensory irritative conditions, and probably act favorably besides in stimulating and regulating the peripheral circulation. Additions of pine-needle extract (1/4 lb. for a full, a few tablespoonfuls for a partial, bath) will often increase the agreeable effect. Baths may be taken every day, or later 2 to 3 times a week; the patientshould stay in at first 15 minutes, afterwards up to an hour. Protracted baths may possibly produce an enfeebling effect upon the general condition, which should, of course, be avoided. For insomnia, the baths will be most effective if taken in the late afternoon.

No indication will usually be found for the use of hot-water baths (100° F.) and other general hot procedures which are, as a rule, more fatiguing. If a cold seems to have produced the palsy—a rather rare occurrence, by the way —diaphoretic treatment may be tried at the outset. In cases of poisoning, too, especially those due to certain metals, one may try to help along the treatment by occasional sweatbaths and packs. Concerning the practical application hydrotherapeutic text-books should be consulted.

Local bog, peat or mud packings (122° F., duration from 1/2 to 2 hours) most likely affect convalescence advantageously during the regenerative stage; at any rate they will have a favorable effect upon symptoms of sensory irritation that still exist.

Douches to produce irritation in sensory nerves, in disturbances of sensibility, may be employed as well as other means which will be considered somewhat later.

Electro-therapeutics is of much importance. It is true that some peripheral paralyses recover without electric treatment, but the omission of an indicated electric treatment in serious cases is always a grave error in judgment. Aside from certain surgical procedures and some necessary measures of general hygiene, all other methods rather than the electro-therapeutic could be omitted. It is, however, not merely a question of employing "electricity" but of employing it *skilfully and by the proper method.* One who can not fulfill this demand should, as a matter of principle, abstain from personally treating peripheral nerve diseases. This may even today be said with a full recognition of the fact that the critical, even the sceptical reaction against the former overrating of electro-therapeutics is justified. Of the many methods recommended for our purpose, we shall especially emphasize the following, approved by long experience. In the present state of our skill and knowledge they will ordinarily suffice.

In case of severe pains in the early stage, stabile anodal galvanization, a classic method of treating neuralgia, which will be described later on, may be

used. The anode is applied at the point of greatest tenderness or in more serious local lesion severing the nerve, in the region of the central stump.

The method just mentioned is a symptomatic one, to be tried occasionally, while to the following two considerable influence on convalescence and the prospect of recovery is justly ascribed.

The stabile cathodal galvanization is applied when the nerve trunk has suffered a lesion, which may be definitely localized, particularly in pressure paralysis, but in other traumatic affections also. The cathode (electrode of about 20 qcm.) is placed over the point of lesion, the inactive (dispersing) anode (about 100 qcm.) on the breast or back. The current is increased and diminished gradually. Current strength: 5–6 (4–8) milliamperes. Duration of treatment: 5–6–10 minutes. Seances to be repeated daily, at least every other day. Treatment should be begun a few days after the paralysis has set in. Reliable experience proves that it is apt to hasten the convalescence (E. Remak). Instead of leaving both electrodes stationary, the active electrode, the cathode, may be moved up and down the course of the nerve trunk to be treated, without, however, removing it from the skin.

The actual galvanic stimulation of the diseased region is the most important treatment in the later stages of the paralysis, and may sometimes be combined with the preceding method, but should never be applied before the third week. The endeavor is to produce muscular contractions through direct stimulation of single muscles. Button-shaped electrodes, whose surface is about 10 qcm. and, if desired, having a current interrupter attachment, are used for this purpose. Generally speaking, the muscle is stimulated with a current of just sufficient strength, afterwards with a somewhat stronger current, applying the cathode as the irritation electrode, or in cases of D. R. when the anode excites greater contractions, applying it preferably. Stimulation is produced by abrupt closing of the current by the interrupter, or by moving the electrode over the muscle to be stimulated. In cases of considerable diminution of the irritability, alternating currents with attached electrodes, so-called voltaic alternating currents, are employed. It is best to let the muscles contract one after the other, and then to begin again with the first. In this way each muscle of the paralyzed area should be stimulated at first, say, 10 times, afterwards from 20 to 40 times. Cylindrical electrodes are also in use with which the diseased area is traversed in a centripetal direction. The inactive large electrode is again placed on the breast or back. Some recommend that it be placed on a proximate part of the member to be treated, or on the nerve trunk whose muscle area is to be treated. If the irritability is not too greatly decreased, some stimulations of the nerve trunk may be added to the galvanization of the muscles. Daily sittings should be held so far as possible.

This method may or must be modified in cases where great sensory

irritability, hyperalgesia, to the electric current exists. All painful fluctuations in the current are to be avoided by a very gradual increase of the current, after the electrode has been attached, and moving the active electrode up and down over the area to be treated, without, however, removing it from the skin. The electrode is taken off only at the termination of the treatment after the current has been gradually diminished.

Further, another method which does not favorably compare with the above and which is based on the so-called refreshing influence of the flow of the galvanic current in penetrating the muscle, may be applied. Two nearly similar, medium sized, flat electrodes are applied in such a way on suitable places of the paralyzed musculature that after closing the current it will penetrate the muscles; the anode on the more tender parts of the skin. The current of 4 to 6 milliamperes is gradually increased and diminished. The duration of the seance, during which the position of the electrodes may be changed by sliding them over the skin, should vary from 5 to 10 minutes.

Just how these methods influence recovery in paralyses, is still problematic; but clinical experience has clearly shown that they prevent, to a certain extent, muscular atrophy and that they promote recovery. Some experimental investigations also give support to this statement.

The faradic current may be used in those cases of paralysis in which the irritability has decreased but little and medium faradic currents suffice to cause contraction of the muscles. They should be stimulated by stroking or rolling of the musculature, or better still by sudden closing of the current. Energetic prolonged tetanization of the muscles can not be advised. However in motor paralysis we decidedly prefer the galvanic treatment. Galvano-faradization is recommended by a number of authorities. Both currents are brought simultaneously to the electrodes, and the currents are graduated in such a way, that each current just suffices by itself to produce contractions. It is always presupposed, of course, that the faradic irritability of the parts concerned has not suffered severely. The faradic current is usually contra-indicated in motor and serious sensory conditions of irritation. In the first stage of paralysis in which genuine irritants are to be avoided, the faradic current should not be employed under any conditions.

The faradic current is particularly to be recommended for the treatment of anæsthesia, if there is no contra-indication for other reasons. A dry, brush-shaped electrode is used, with which the anæsthetic area is stroked or, moving it from place to place, tapped with a current just strong enough to excite the sensibility. By the use of a cylindrical or movable button electrode in galvano-faradization it is possible to do justice at one and the same time to the indication of the muscle irritability and the irritation in sensibility.

The electric treatment should generally be continued until convalescence is established. Treatment of a few weeks in grave paralysis will not be effec-

tive. It will be clearly seen in many cases that in every stage of the convalescence electric treatment acts beneficially, while the omission of it will retard the progress of recovery. Sometimes, in inveterate cases, improvement sets in only after the electric treatment has been begun. The ameliorating effect which it frequently and directly produces on function, is especially adapted to animate the spirits of the patient with all its consequences. Experience proves that patients hardly follow as faithfully any other method for such a long time as they will electro-therapeutics, even if the progress of recovery be but slow.

This treatment should be tried even in quite inveterate cases. As long as the musculature is still capable of being electrically irritated and the nerve conductivity is not wholly destroyed, even if only a prospect for the regeneration of the nerve exists, there is still hope for success.

In the stage in which absolute rest does not seem necessary any longer, *massage* materially assists electric treatment of the muscles. Exact procedure based upon anatomy and performed best according to Hoffa's excellent instructions[1] is indispensable.

Besides effleurage, gentle, later vigorous kneading and careful percussion may be instituted; the more energetic manipulations should be used only in the stage of convalescence. Even if massage of the muscles may in a few exceptional cases be entrusted to a specially trustworthy unprofessional masseur, massage of the nerve trunk should be attempted only by an experienced professional hand and then only in a few exceptional cases. The very great liability of injuring this delicate structure should always be kept in mind.

Medical gymnastics in its widest sense, is absolutely necessary in a cure of peripheral motor paralysis. As soon as mobility returns, each injured muscle should be exercised systematically. One should never depend on the patient's using his muscles of his own volition because he fails to do so only too often, or if he really uses them, he does so usually to an insufficient degree.

But even in the stage of complete paralysis, as early as the fundamental principles already laid down will permit, gymnastics, at first only carefully executed passive movements, should be instituted. By this means the development of contractures is prevented. This danger should be thought of in the beginning also, particularly in reference to the position of the diseased parts. In a peroneus paralysis a pes equinus position is not to be tolerated; in a radialis paralysis the hand is not to be left flexed. Proper support and bandages should keep the member in a medial position. Above all, passive movements promote the return of active motility. The patient should from the beginning endeavor actively to assist the passive motion. The moving hand perceives immediately when these endeavors begin to be successful. Then the passive motion is replaced by passive-active gymnastics. The dis-

[1] His "technic of massage" (Stuttgart, 1903) is, in its technical suggestions, authoritative.

eased member requires aid for some time yet, if it is to utilize the existing minimum of strength gymnastically. This support is rendered by the hand of the physician or his assistant, who have so far moved the member only passively. In many cases an unaffected hand of the patient may render the assistance, in suitable cases, by means of a skilfully attached strap. Thus if a part is to be dorsally flexed a strap may be attached to it and worked by the patient. An excellent means of aiding the first attempts in active gymnastics is afforded by warm baths. The laws of physics enable the patient to execute motions independently in the bath which he is unable to make otherwise (kinetotherapeutic baths) and this is a particularly important point indicating the bath treatment (Goldscheider).

If apparatus for so-called semi-active gymnastics is at one's disposal, apparatus which mechanically assists the performance of the patient and enables him to make movements with the expenditure of but little strength, like the Krukenberg pendulum apparatus and similar contrivances, it may be used.

With further progress toward recovery the real movement exercises and finally the various forms of resistance gymnastics are reached.

As a general rule for gymnastic therapy it may be stated, that here as everywhere, the patients should exercise frequently, but not long at a time; that they should never be tired out or too much exhausted by it.

Surgical treatment, in many cases the conditio sine qua non of recovery, is discussed separately in this work. We shall therefore simply suggest here the general possibility of its application.

When a nerve is functionally injured by surrounding scar tissue, by pressure of masses of callus, or, in a similar manner, neurolysis, notwithstanding the fact that in some cases a spontaneous recovery may obtain, should, as a rule, be used promptly as it sometimes restores the nerve conductivity directly. A completely divided nerve should be sewn, if possible, primarily, as restoration without the reunion of the nerve-ends, despite some sporadic, anomalous cases, not as yet explained, can not be expected. If any doubt exists as to the necessity of a suture, it is in most cases advisable to look for the nerve operatively, and to decide then whether or not to suture it. In inveterate cases, where the course has demonstrated that an amelioration is not to be expected without surgical intervention, the different methods of even secondary nerve suture promise success. If this is impracticable the advisability of nerve grafting will have to be considered.

Of course in all these cases, with but few exceptions which are difficult to explain, a long time will elapse before functional restoration takes place.

If the morbid process has entirely ceased, and it is quite obvious that a change, either for better or for worse, will not set in, and if the procedures for the restoration of innervation are no longer a point of issue, orthopedics are in order. Attempts should be made to give the paralyzed member support

by means of immobilization apparatus, by arthrodesis, by operative shortening of relaxed tendons. Contractures are relaxed by tenotomy or by plastic operations on tendons; unimpaired muscles, by transplantation of tendons, are made to serve as substitutes for muscles whose functions have been lost.

In many cases such measures will produce extraordinary results. But one must not expect too much from orthopedic operations. If the patient is used to an old defect, and endures it without considerable impairment of his well-being and his capabilities, it would mostly be well to omit surgical operations, the result of which is never quite certain.

Finally let it be particularly emphasized that besides operative procedures the other methods of treatment should never be neglected.

PARALYSIS OF INDIVIDUAL PERIPHERAL NERVES

Although in the following the clinical picture of diseases of individual peripheral nerves is to be discussed, it will not be done with exclusive regard for the clinical group of single diseases. We shall keep in mind that a great part of the paralyses to be treated here, appear as partial symptoms also in polyneuritis.

A. The Cranial Nerves

1. The Olfactory Nerve

It is obvious that the olfactory nerve may, in a number of intra-cranial diseases, be affected unilaterally or bilaterally. In tabes it may atrophy.

Independent isolated affections of the olfactory nerve are rare. Only one type is of some practical interest. After trauma of the skull, anosmia, which can be ascribed to the mechanical destruction of the olfactory filaments, may occur. The loss of the sense of smell may mean an impairment in the ability to make a living, especially in those occupations in which the tasting and "smelling" of food or drink is essential, and in others in which the organ of smell is necessary to protect one's self from dangers, such as poisonous gases (chemical factories, etc.).

Destruction of the olfactory nerves results in loss of smell. At the same time, however, there is involved great impairment of the ability for what is colloquially termed "tasting." The ability to taste is in organic anosmia restricted to the elementary taste qualities (sweet, sour, etc.). On this the differential diagnosis in regard to hysteric anosmia may be based. One affected with organic anosmia, despite the preservation of the organs of taste, lacks the ability to distinguish all those articles by the "taste," for whose recognition, unknown to most people, the co-operation of the sense of smell is necessary. If this ability is preserved in spite of anosmia, it points unquestionably against an organic foundation.

It must be remembered that by far the most frequent cause of anosmia is nasal disease, and that it must be carefully excluded before nervous anosmia is diagnosed. In obstruction of the posterior nares the sense of smell, in subserving taste, is much diminished. Patients complain about disturbances of taste, even though the sensitiveness of the organ of taste for the elementary qualities is normal and though odors are recognized by the nose (gustatory anosmia).

Congenital defects of the olfactory nerve also occur. The fundamental factors of senile anosmia, and of that which sometimes remains after nasal disease, are still unknown.

2. The Optic Nerve

The diseases of the optic nerve of interest to the neurologist are those which occur most often as part of the symptomatology of certain cerebral and spinal diseases. Isolated diseases of the optic nerve, as a rule, fall to the lot of the eye specialist. Nevertheless, especially in the interest of differential diagnosis, a survey of the general field, which we shall give in the following paragraphs, is necessary to the neurologist.

The **optic nerve,** in the morphologic embryological sense, is not a peripheral nerve, but a projected portion of the brain. Of especial significance for an understanding of its diseases is the fact that the fibres serving to give the keenest central vision, belonging to the yellow spot (macula lutea), form in the nerve trunk a large separate bundle. This papillo-macular bundle first occupies, near the bulbus, a peripheral position corresponding to the temporal border of the disc, while further on it runs in the axial part of the transverse section.

The chief **symptom** of disease of the optic nerve is clearly disturbance of vision of various forms, which, however, differ from those occurring in affections of the chiasm, the optic tract, and the more central parts of the visual tract, which are distinguished by their peculiar hemianopic character, and which disease of the optic nerve does not produce. (Concerning hemianopsia, cf. the chapter on brain diseases.)

In serious disease of an optic nerve the eye concerned, almost without exception, does not react to light by contraction of the pupil. Of more theoretical interest than practical importance is the fact that in very rare cases, where total blindness resulted from atrophy of the optic nerve, the optic nerve concerned was said to have transmitted pupillary light reflexes. According to this it would seem that special fibres subserving the reaction to light could withstand, on occasion, the morbid processes longer than the tracts destined for vision, though even in these cases the pupillary light reaction never was totally undisturbed.

In most diseases of the optic nerve ophthalmoscopic changes of a characteristic nature are known to take place in the disc of the optic nerve.

For various reasons a *classification of optic-nerve diseases* is very difficult. Concerning the pathological anatomy there is no unanimity among the most experienced investigators, and still less clear in many respects is the pathogenesis. The same visible changes in the fundus are occasioned by various conditions, and, on the other hand, the same injury may occasion quite different anatomic and ophthalmoscopic pictures. Finally, the ophthalmoscopic findings change considerably in various stages of the same process.

The following classification will suffice for all practical purposes, though it is necessarily lacking in unity.

(*a*) *Primary* (*Idiopathic*) *Atrophy*

Idiopathic atrophy is diagnostically a very important symptom, because it often appears very early, of tabetic and paralytic diseases. It will be treated more minutely in its relation to those diseases in the proper place. Clinically it is characterized, besides the ophthalmoscopic findings of an always bilateral, simple diffuse atrophy, by a more or less severe, usually irregular, but concentric contraction of the field of vision. The sense of color suffers likewise (at first for red and green), and so does central vision. The fibres of the optic nerve disappear, and likewise very early their cells of origin in the retina. *Simultaneously with the atrophy*, there gradually develops a disturbance in sight, which usually progresses till total blindness results.

The atrophy of the optic nerve in glaucoma, which is of purely oculistic interest, may always be distinguished definitely from tabo-paralytic atrophy by ophthalmoscopic examination.

The atrophy of the optic nerve in the Tay-Sachs amaurotic family idiocy is probably the result of a serious disease of the retina. The ophthalmoscopic picture, the circumstances and the symptoms accompanying its appearance are so characteristic that it never presents any difficulty in differential diagnosis.

The secondary atrophy after papillitis (neuritic atrophy) will be discussed in the following paragraphs. It, too, is differentiated from "simple" atrophy by the peculiar ophthalmoscopic findings, which will in most cases enable its recognition and differentiation, especially in the early stages in which the differential diagnosis of the affection of the optic nerve is of prime importance for a decision as to the fundamental morbid condition.

The primary tabetic and paralytic atrophy may most easily be confounded with certain *secondary simple atrophies*, which appear most frequently in connection with retrobulbar processes of the most varied nature. They are treated of under (c) and (d), where reference is made to the peculiar characteristics of the different forms in this group.

(*b*) *The Forms Occurring under the Aspect of Papillitis, Especially Choked Disc and Primary Neuro-papillitis, Neuritis Optica Intraocularis*

In its incipient stage the prognosis of a papillitis may present considerable difficulties. When the physiological tortuosity of the vessels of the retina is strongly marked, and the disc is hyperemic, as is habitually the case in hypermetropia, it is really a case of pseudo-neuritis optica. The concentric wrinkling of the retina in the neighborhood of the disc, which is occasionally observed in the beginning of true papillitis, may make a differential diagnosis possible.

1. **Choked disc** occurs as a result of space contracting intra-cranial processes, or, to be more precise, of such conditions as prevent the free flow of the venous and lymphatic circulations in the cavity of the skull. This causes trouble in the area of one or usually of both optic nerves, which is the immediate cause of the development of choked disc. Disease processes in the orbit, too, may in rare cases lead to analogous, though, of course, unilateral, alterations. Choked disc will be treated in more detail with the different intra-cranial diseases especially with brain tumor. From choked disc it is not always easy by means of an ophthalmoscope clearly to distinguish the next affection.

2. **Primary Neuritis Optica Intraocularis.**—The histological and pathogenic comprehension of choked disc in its more important manifestations is not yet very definite, and this holds true, perhaps even to a higher degree, in the case of neuritis optica. In the latter, it is usually a case of inflammatory processes, while choked disc, at least in the early stage, shows only congestive symptoms, especially edema. As a primary independent disease, neuritis optica has been traced back to all sorts of sources, frequently on very questionable grounds. Its occurrence in connection with various infectious diseases, intoxications (lead), and with intra-cranial infectious inflammations, is certain. It is supposed that it may be directly produced by anemic conditions and severe hemorrhage, Worthy of mention is a hereditary form, usually appearing at puberty in males, and being transmitted through the female line.

3. **Papillitis in inflammation of the retina** (albuminuric retinitis et al.), changes of the disc in thrombosis of the central vein and in embolism of the central artery, have a decided differential diagnostic interest for the neurologist.

All forms of papillitis are accompanied by more or less serious disturbances in vision. These may recover or result in so-called secondary neuritic atrophy, which ophthalmoscopically is characterized by certain peculiarities.

In choked disc, vision more frequently remains normal for a considerable time.

(*c*) *Diseases Beginning in the Retrobulbar Region* (*Retrobulbar Neuritis*)

In cases of this group, the disorder begins with a destructive influence acting upon that section of the optic nerve which is retrobulbar. Hence the first clinical symptoms are disturbances in vision, sometimes total blindness, sometimes peripheral defects in the field of vision, sometimes disturbance of central vision without impairment of the peripheral parts of the field of vision, so-called central scotoma, which indicate an isolated disease of the papillo-macular bundle, for which, according to the latest investigations, an anatomic predisposition to peculiarities in the supply of vessels of this area, is apparently to be held responsible (Birch Hirschfeld).

According to the place, nature and mode of attack of the influencing injuries, ophthalmoscopic changes occur in a few cases relatively early, as neuritis optica, which may be caused by a descending inflammation, or choked disc. In such cases the disorder may terminate in atrophy of the disc.

In many cases, however, the visible changes of the papilla of the optic nerve occur only later, when, according to the extent and gravity of the disease, it appears as a more or less marked, usually only partial, especially often temporary, simple atrophic discoloration.

Retrobulbar optic-nerve disease is as a rule unilateral.

From the facts stated, the necessary general basis for differential diagnosis becomes apparent.

Not considering the rare focal diseases, such inflammatory processes as occur in myelitis and meningitis, and the metastatic abscesses of the optic nerve which are sometimes observed, the following diseases must be considered chiefly.

(1) Diseases due to pressure. The effect of long continued pressure on the nerve trunk, occurring in tumors in the immediate vicinity, in aneurisms of the internal carotid or ophthalmic artery, in old age perhaps even in simple arterio-sclerosis of these vessels, usually is simple atrophy.

Constant pressure on the chiasm (tumors of the hypophysis) or the optic tract may also lead to descending atrophy. Sight disturbances in these cases generally are hemianopic; many diseases of the chiasm even lead to total blindnesss.

In cases of very slowly increasing pressure, it may occur that disturbances in sight will not be noticed until visible atrophy has set in, which is often easily confounded with simple atrophy.

(2) The tumors of the optic nerve itself, of which many primary and metastatic forms occur. As a rule they lead to papillitic processes.

(3) Other orbital diseases of all kinds may encroach upon and affect the optic nerve.

(4) The affections of the optic nerve in diseases of the posterior accessory

cavities of the nose, of the sphenoid and ethmoid cavities, which stand in close anatomic relation to the optic trunk.

(5) Malformations of the skull, especially the so-called tower-shaped head, frequently, even in infancy, lead to disease of the optic nerve.

(6) Injuries of the optic nerve caused directly by gunshot or knife wounds, and more frequently by fracture of the skull, show naturally all degrees of lesion up to the complete rupture of the nerve. In the beginning the disc may be hyperæmic or edematous; in ruptures of the central vessels it presents the appearance of grave anæmia. Very often the ophthalmoscopical picture is at first quite normal. The outcome is always an atrophy, usually simple, corresponding in degree to the injury. The atrophy of the optic nerve which is sometimes observed in small children after delivery by means of forceps, may also be included here.

In some rare instances, after cranial trauma, bilateral atrophy of the optic nerves has gradually developed, which clinically has usually taken the same course as primary tabo-paralytic atrophy.

(7) Diseases of the optic nerve in multiple sclerosis.

Multiple sclerosis very frequently leads to the development of focal lesions in one or both optic nerves. The peculiarity of this disease to leave the axon intact for a comparatively long time, explains some singularities in the clinical picture of this disease of the optic nerve. Disturbances of vision may be entirely wanting, even in cases in which an atrophy ophthalmoscopically proven, most commonly with its characteristic picture of temporal pallor, exists. In the course of the further development of the malady, slight disturbances in vision may still set in. They rarely, if ever, develop to such a degree as to result in marked diminution of vision, let alone blindness.

Not infrequently acute disturbances in vision are observed in the early stage of the disease, mostly of one eye, as a rule in the form of central scotoma, occasionally with transient papillitic changes, and in exceptional cases even choked disc. These sight disturbances are generally more or less completely recovered from. They are probably coincident with the early stage of a sclerotic focus in the optic nerve.

(*d*) *Toxic Amblyopias and a Few Related Forms of Acute Blindness Originating Peripherally*

That many different intoxications may lead to the development of papillitis has already been mentioned. The toxic amblyopias occupy a unique position with respect to these affections. Under this name acute and chronic diseases are grouped together.

The toxic amblyopia of chronic development, observed most frequently in chronic tobacco and alcohol poisoning, may be considered as one of the retrobulbar diseases which we have just discussed, though it is as a rule

bilateral. Concerning its pathological anatomy and pathogenesis, nothing conclusive is known, but the clinical picture indicates that the papillo-macular bundle is chiefly affected. The disturbance in vision is a characteristic scotoma, first for red and green, later on for white, while the field of vision otherwise remains normal. In the later stages, a slight pallor, especially of the temporal part of the optic nerve head may set in. Mistaking it for incipient tabetic atrophy can be easily avoided by observing carefully the characteristic functional disturbances. The prognosis is relatively favorable; that of the tabetic atrophy, unfavorable.

Bilateral toxic amblyopias and amauroses of peripheral origin setting in acutely occur in many acute poisonings, among which, those due to quinine,[1] male fern, and methyl alcohol should be mentioned. They have been also observed to result from prolonged use of atoxyl. Very similar clinical pictures may be produced by severe hemorrhage, and, after employment of X rays or other rays of somewhat similar character. They are occasioned by grave, acute diseases of the ganglionic retinal nerve cells, and often of the fibres themselves. A partial restoration is possible but the usual termination is a more or less complete atrophy of the optic nerve.

What produces the grave disease in the optic apparatus in these and other cases in which general injurious influences are at work, is not known.

The **therapeutics** of the diseases of the optic nerves must proceed according to fundamental principles. In cases of atrophy, in which amelioration, or at least a cessation of the process, does not seem impossible, galvanic treatment should be tried, though the recent optimistic reports concerning the success of such treatment need to be more fully verified.[2] The anode is placed upon the closed eyelid, the cathode, which should be larger, upon the neck. Strong currents, carefully increased and diminished, and protracted seances (15 minutes and longer) are recommended. Sometimes long continued treatment with strychnine appears to be useful.

3. Nerves of the Eye Muscles. Oculomotor, Trochlear and Abducens Nerves

The palsies resulting from disease of the motor nerves of the eye, in a great number of cases can, practically not, be distinguished with certainty from those resulting from intra-cerebral injury of the same peripheral motor neuron. Even theoretically, in a large group of cases, it can not be authoritatively stated whether they may be considered as peripheral nerve disease or not.

Palsies due to progressive nuclear (spino-bulbar) muscular atrophy, cases of polioencephalitis hæmorrhagica superior, probably most cases of congenital palsies caused by imperfect development of the nuclei which often assume the character of supranuclear paralysis, and finally the paraly-

[1] The minimum dose that has occasioned disturbances in vision, is 5 grm. in 30 hours.

[2] Cf. Mann, Zeitschrift für diätet. und physic. Therapie, volume VIII.

ses occurring in focal disease of the crusta, either injuring the nuclei themselves or affecting the intra-cerebral root-bundle sympathetically (fascicular paralyses), all these can be traced back to nuclear or at least to intra-cerebral injuries or diseases.

In contrast to these forms, which are mentioned here only for the purpose of differential diagnosis, stand others which most certainly are peripheral nerve diseases, i. e., the large group of basal and orbital paralyses. Among the diseases which cause paralysis by lesion of the nerves, acute and chronic meningitis from whatever cause, and especially syphilitic chronic basal meningitis, must be first mentioned. Injury to the nerves caused by the pressure of tumors must also be considered. These tumors may be situated in the meninges themselves, though more frequently those of the base of the skull (sarcomata), and above all, tumors of the brain are the cause of the paralyses through pressure. It is very important to notice, that tumors of each brain region may occasionally cause ocular paralysis. Tumors in the region of the posterior cranial fossæ most frequently injure the abducens; those of the middle cranial fossæ, especially of the temporal lobes, may lead to grave and extensive ophthalmoplegia through pressure on the nerve trunks, which are closely interwoven at the base of the brain, as is the case with the tumors of the frontal region when they spread towards the superior orbital fissure. In the cases named, the mechanism of the pressure effect is clearly understood. But sometimes there are cases where it is very difficult to understand the relation between a brain tumor located at some distance from the base, and some coincident forms of slighter paralyses in the eye muscles. Neoplasms very rarely directly involve the nerves. It is exceedingly seldom that tumors arise on the nerves themselves (primary and metastatic).

Besides morbid growths, arterio-sclerotic changes in vessels, aneurisms of the internal carotid and other arteries, can injure the nerve trunks through pressure.

Like tumors of the temporal lobes, abscesses of the same region affect the nerves of the base.

Thrombosis of the cerebral sinuses, particularly of the sinus cavernosus, frequently lead to paralysis of the eye muscles.

Of diseases of the orbit, it is mainly inflammation and tumors or tumor-like structures that play an ætiological rôle.

Of the traumatic palsies, those resulting from fracture of the base of the skull are of the greatest interest.

Trauma and syphilis may likewise in rare cases lead to paralyses from intra-cerebral injury or disease. The trauma may cause hemorrhages in the nuclear region, while syphilis is likely to induce chronic progressive disease of the nuclei.

Those cases that now and then are observed in connection with ophthalmic herpes zoster are probably also of neuritic origin.

For an exceedingly large group of paralyses, as has been said before, no definite classification which would serve for all cases can be made, since the known pathologic-anatomical findings vary, at least enough to make different explanations possible. This is the case with paralysis of the eye muscles in tabes and paresis, in multiple sclerosis, and in the cases connected with infectious and toxic diseases. Among the infectious diseases, diphtheria and influenza stand in the front rank (cf. also chapter on polyneuritis). Among the toxic processes, sausage poisoning (botulismus) and lead poisoning deserve to be mentioned. The cases observed in Basedow's disease and diabetes most likely depend on autointoxication. How in all these cases there results a localized affection of special eye muscles is entirely unknown. Very uncertain is the ætiological significance of catching cold or being chilled, so often affirmed. In many cases all discoverable causes are lacking.

Clinical Conditions and the Course of the Disease.

Paralyses of the eye muscles appear, as is evident from the ætiological discussion, as component symptoms of the most varied conditions. However, they appear also as an independent disease.

In either case the affection may be restricted to one of the cranial nerves or to a nerve pair. Isolated oculomotor and abducens palsies are frequent; isolated trochlear palsies rare. The trochlear is more frequently affected in combination with other ocular nerves.

In oculomotor affections, total paralysis and paresis affecting all branches of the nerve must be distinguished from partial functional disturbances, restricted to a few single muscles of its innervation area.

In total oculomotor paralysis the mobility of the eye is practically gone. As the abducens and trochlearis are functionally unimpaired, the eye deviates considerably outward and a trifle downward. Every attempt to innervate motorially the ball is apt to increase this deviation. The eye is diverted still further into the outer canthus, while, because the trochlearis still acts, the upper pole of the vertical meridian of the cornea is at the same time turned slightly inward. The increase in deviation due to an attempt at innervation, no matter how directed, must be looked on as a so-called compensation movement and is explained thus: that an innervation current flows off into the open channels, since its own are closed. The upper eyelid is dropped and can be raised only passively. The pupil is more than medium wide, the contraction reactions are lacking, the accommodation is paralyzed. Often there co-exists a slight degree of exophthalmus. The eye, since its muscular fixedness is lacking, usually almost protrudes from the orbit.

Partial oculomotor paralysis may affect each muscle separately or an irregular group of muscles. To the most frequent occurrences of this nature belongs isolated paralysis of the levator palpebræ (ptosis). Important

typical combinations are the so-called external oculomotor paralyses, in which only the external branches of the nerve are paralyzed,[1] and the contrasting ophthalmoplegia interna, in which the paralysis is confined to the internal muscles of the eye. It is of special importance to know the conditions in convalescence of the latter, and of great practical interest, too. As a rule, first the palsy of the ciliary muscle decreases, and the accommodation naturally returns, while the pupil does not contract either to light rays or by convergence. (A condition known as pupillary immobility, "Pupillenstarre.") In the further course the convergence reaction returns before the light reaction, so that in this stage it is easy to confuse this condition with that of purely reflex pupillary immobility, which, diagnostically, is so extremely significant. For the differential diagnosis it is important first to note that ophthalmoplegia interna frequently occurs unilaterally, while the true Argyll-Robertson phenomenon, on the other hand, occurs unilaterally only in exceptional cases. In residual ophthalmoplegia interna, the pupil as a rule is of an average or slightly larger width, while the reflex pupillary immobility often combines with miosis. The most important point is, that in reflex pupillary immobility the convergence reaction always remains absolutely normal. In the spurious form slight residua of a disturbance can always be found, even in regard to the convergence reaction.

Besides thus grouping these cases according to the extension of the paralytic symptoms in the different eye muscles, a further classification according to their varying courses is necessary. We know of cases setting in acutely, frequently comparatively mild, even transient forms; further of chronic progressive, and finally of relapsing and periodically reappearing cases.

Whether a paralysis starts acutely, whether its course is mild and regressive or continuing, is, of course, dependent upon the nature of the influence, and the reparability of the causal anatomic process. Those ("rheumatic") paralyses which some writers have traced to colds, are famed as mild cases, and the paralyses associated with herpes zoster ophthalmicus ought to permit of a favorable prognosis. The palsies occurring in tabes and multiple sclerosis, after lasting a short time, may disappear completely and forever.

On the other hand, however, these very ophthalmoplegias of tabes, general paralysis and multiple sclerosis may take a chronic progressive course. Several others, certainly the nuclear forms of the disease taking a like course, need not be specially discussed here.

The recurring form of paralysis of the eye muscles occurs, too, in tabes and paresis. A second group of recurring paralyses, which actually take a periodic course, and which are almost invariably restricted to an oculomotorius, may, according to anatomic findings, have their origin in pressure of tumors on the nerve trunk, or in chronic meningitic and arterio-sclerotic

[1] Strictly speaking, ophthalmoplegia externa totalis should be assumed only when all external eye muscles including, of course, those innervated by the abducens and trochlearis, are affected.

processes taking place in the immediate vicinity of the nerve and injuring it. The duration of the exacerbations, and the periods of remission and intermission will naturally be very different in the various cases. At the time of aggravation, a headache on the side affected manifests itself, sometimes accompanied by vomiting, in consequence of which, it has become common to speak of it as "migraine ophthalmoplegique" from a certain superficial resemblance in clinical pictures. These conditions, nevertheless, have nothing in common with genuine migraine. However, it is said that even in genuine migraine some transient attacks of eye-muscle palsy occur.

Diagnosis.—The first problem, though by no means always the easiest one, is to find out definitely whether a paralysis of the eye muscles exists at all, and what muscles it affects. For this the reader is referred to the discussion in the general section dealing with diagnosis.

In the second place we must try to discover whether it is a supranuclear or a peripheral paralysis proper, using that term in its widest sense. Finally, and of most significance to us here, we are confronted by the most difficult, only too often insoluble, question: which portion of the peripheral neuron is the primarily diseased part—is it the nucleus, the intra-cerebral root-fibres, the intra-cranial or the orbital part of the peripheral nerve?

At first we must say a few words concerning the differentiation of disease of the peripheral neurons from the supranuclear or looking-paralysis (Blicklahmüng), a problem the solution of which needs special tests.

Generally speaking, in diseases of the peripheral neuron only, it is a matter of a true paralysis of the *muscles*, while in supranuclear lesions only certain *functions* of the muscles disappear. Only in peripheral paralysis is the muscle equally cut off from all inflowing nervous impulses. A supranuclear focus, on the other hand, interrupts only those pathways which convey to a certain group of synergists the impulses for a certain common function, so that the latter is suspended, while the same muscles may remain normal for other functions, and open to inflowing innervation from other sources. Thus the "looking motion" (Blickbewegung) towards one side may be palsied, while the co-affected internus is active in convergent motion. A supranuclear lesion, however, may also be restricted to the fibres which convey the impulse to one of the synergists. In that case, only this one muscle is affected, but only for the function concerned, without therefore being incapacitated for other functions. In such cases, just as in peripheral paralysis, disturbances in equilibrium and double images manifest themselves with each innervation of the synergism, which in one of its parts is injured. Decisive for the differential diagnosis as to the supranuclear seat is, therefore, the proof that the paralytic symptoms appear only in certain functions, while the same muscles for other functions remain normal. One must consequently make different tests. First, the patient is commanded to look in this or that direction, or to fix with his eye an object located

within the peripheral part of his field of vision. Besides these "commanded" movements, one should test the so-called reflex eye movements. In the first place, the patient is made to follow with his eyes a moving object—which he has previously fixed—in all directions; and secondly, he is made to hold with his eyes a fixed object located in the primary direction of his vision, while his head is passively turned in all directions. In the particular case of a paralysis of an internus for the lateral looking movements, his convergence function should also be examined, and vice versa. In the majority of cases, these methods will suffice to determine the above mentioned characteristics of a supranuclear paralysis. In other cases different criteria are required. It is conceivable that because of complicated lesions the reflex as well as the "commanded" movements may be synchronously suspended, while the peripheral neuron is not diseased. In that case, the strictly symmetrical character of a paralysis may serve as evidence of its supranuclear origin. An absolutely symmetrical paralysis of two or more synergists, in which no disturbance in the muscular equilibrium could be detected, does not occur, aside from cases of total paralysis, in peripheral diseases. It is evident, however, from the above, that the converse need not be necessarily true, i. e., every paralysis that affects only one synergist, or several unevenly, is not by reason of this a peripheral palsy.

If these criterions be heeded, the neuron diagnosis will in most cases be successful. To decide more exactly in what part of the peripheral neuron the seat of the lesion is to be looked for, and particularly to decide whether it is a question of a disease of the peripheral nerve, the following points deserve consideration. In an isolated disease of the abducens or trochlear, this decision can naturally be made only in those cases, where accompanying symptoms render possible the diagnosis of the fundamental anatomic process and its seat. Even for oculomotor paralysis this is frequently the surest and often the only practicable way.

It was assumed formerly, that, excluding cases due to orbital disease, the paralyses of single muscles supplied by the third nerve, were all caused by nuclear disease. At present we know that disease of the nerve trunk may lead only to paralysis of one or a few muscles, and, indeed, that this occurs not infrequently. Isolated ptosis can, in numerous cases, be traced to disease of the nerve trunk. Partial disease of the trunk, for example scattered foci of disease, have been proved anatomically in a number of such cases. The picture of external oculomotor paralysis may surely be brought about in this way. There is no accurate explanation for it. It is doubtful whether a unilateral external oculomotor paralysis may result from nuclear disease. The fibres proceeding from the nuclei of both oculomotor nerves are subjected to a partial crossing, so that each nerve is composed of crossed and uncrossed fibres. Just how both are distributed

to the various muscles, is a question which has not yet been finally settled. The clinical symptoms which follow the destruction of one oculomotor nucleus are not as yet known in detail. This is responsible in part for the great difficulties confronting us in the task with which we are here concerned. Only this may be said with certainty, that a total paralysis of all branches of the oculomotorius will have to be localized distally from the partial crossing of the fibres, in the majority of cases, basally.

Our knowledge concerning the localization of ophthalmoplegia interna is also very incomplete. It is pointed out that a connection of an ophthalmoplegia interna with a paralysis of the inferior oblique muscle indicates orbital disease, since the short root of the ciliary ganglion which contains the oculomotor fibres of the inner eye muscle originates from the nerve branch destined for the inferior oblique.

Unfortunately we know of no other pathognomonic classification of the symptoms, which would be even partially accurate for certain places in the peripheral pathway. When several different eye muscles are affected simultaneously, it denotes with certainty a basal or orbital seat. The cases taking a chronic progressive course most frequently are the result of nuclear disease; those taking an acute and mild course, and the relapsing and periodic cases, are generally due to peripheral nerve disturbance.

The differentiation of certain myopathic ophthalmoplegias from neuropathic forms is frequently possible only by taking into consideration the disease picture as a whole. (Cf. especially the chapter on myasthenia.) Ophthalmoplegia interna, occasioned by the effect of a blow on the eyeball, is probably myopathic.

The **prognosis** of an ocular paralysis should always be made with great reserve. Above everything else the fundamental disease must be considered. Even in those cases in which it is possible to diagnose the pathologic-anatomic process and to recognize it as remediable, it will not always be certain whether the changes this has occasioned in the nerve are likewise adjustable. Concerning their nature and gravity a decision is frequently impossible. But only the inveterate cases and those taking a chronic progressive course are hopeless from the beginning. A favorable prognosis is possible in the acute cases, occurring in connection with infectious diseases and with a number of toxic diseases (botulism). Some other important points for the prognosis have been mentioned in preceding paragraphs.

Therapy.—There can really be no question of special therapeutics for paralyses of the eye muscles. Whenever possible the treatment should be aimed at the fundamental disease. In all other respects one should proceed according to principles laid down for the treatment of paralyses in general. The use of the stabile cathode on the closed eyelids of the diseased eye should be tried. The inactive electrode is placed on the neck. From an authoritative source comes the suggestion that a few sweat baths be prescribed in

those acute cases, which presumably are due to an infectious cause or exposure to cold.

Later, exercises to improve vision, and in the residual stage oculistic and especially operative measures for the adjustment of the disturbances of vision, will have to be considered.

4. The Trigeminal Nerve

The trigeminal nerve arises, like a spinal nerve, from the central organ with an anterior motor and a posterior sensory root. The posterior root enters the Gasserian ganglion. Distally from it the motor root unites with the third of the sensory nerve branches proceeding from the ganglion; the first two of these nerve branches consequently remain purely sensory. Besides combined disease of both roots, there may be disease of the sensory part alone. Cases of purely motor trigeminal paralysis (masticatory facial paralysis) are also on record. Besides the affections of the chief trunk, paralyses of single branches have been observed.

Trigeminal paralysis usually makes its appearance as a partial symptom of other disease pictures; it very rarely occurs isolated.

Its most frequent cause is basal disease. In basal syphilis, in tumors of the base of the brain or skull, and after fracture of the skull, one may occasionally observe isolated trigeminal paralyses, though even in these cases it is seen more frequently in conjunction with palsies of other cranial nerves.

Of diseases of single branches, that of the ramus ophthalmicus, due to morbid changes in the superior orbital fissure or inside the orbit, is observed most commonly.

In a later chapter it will be shown that neuralgia may be followed by symptoms of lost function in single trigeminal branches.

During the last few years, extirpation of the Gasserian ganglion in grave cases of neuralgia has furnished repeatedly an opportunity for studying the symptoms of trigeminal paralysis.

A lesion of the motor part gives rise to paralysis of the muscles of mastication. It may be both seen and felt how the temporal and masseter muscles on the affected side fail to contract, either wholly or partially, when chewing and biting, and how they gradually atrophy. The lower jaw reflex is also abolished on that side. The lower jaw, as a result of the paralysis of the pterygoids, can not be moved towards the healthy side, it tends rather, during chewing, to deviate towards the affected side. In bilateral paralysis the jaw hangs down loose; it can be lifted only with the hand and an habitual dislocation may develop.

Paralyses of the other muscles supplied by the motor trigeminus (mylohyoid, anterior belly of the digastric, tensor tympani and tensor veli palati) do not occasion very significant symptoms.

The disturbances of sensation affect, according to the seat of the lesion, the whole innervation area or that of single branches. For a better under-

standing of the clinical pictures and particularly of the rather far reaching adjustability or possibility of compensation, it is exceedingly important to know, that the innervation areas of the right and left nerves, the areas of the single branches among themselves, and finally the areas of the trigeminus and that of the neighboring cervical nerves may overlap one another extensively.

It should further be remembered, that the innervation area of the trigeminus is not confined to the skin of the face. Of those parts that can be examined, the first branch supplies the cornea and conjunctiva as well as the anterior and upper part of the nasal cavity; the second, the remaining part of the nasal cavity, the upper jaw and the mucous membrane of the palate and upper lip; the third, the lower jaw and the mucous membrane of the cheek, the tongue and the lower lip.

Besides the sensory symptoms of loss of function, pains frequently occur, which, especially in compression of the nerve trunk, may be neuralgic and be projected into the peripheral extension area.

Together with the conduction of sensibility, the conjunctival and corneal as well as the sneezing and palatal reflexes, which are mediated by the sensory trigeminus path, are interfered with. The functions of the lachrymal and salivary glands, especially those of the former, suffer from the loss of a great part of the reflex irritations, that stimulate their secretion. As a result of this and of other disturbances in the region of the smaller mucous glands, there occurs a certain dryness of the conjunctiva and the mucous membrane of the nose, which may even in some cases, it is said, impair the sense of smell. The real secretory nerves of the glands mentioned are not, however, generally affected. A glance at the diagram (Fig. 28) will show that these fibres do indeed mingle with branches of the trigeminus that are directed towards the periphery and that in the last part of their course they take the same course as these branches. Lesion of them in the trigeminus area would therefore be possible only if those terminal branches should be subjected to an injury. As a rule, then, a true paralysis of glandular secretion does not occur, and it may be expected that the psychic emotional secretion especially will go on quite undisturbed.

Fig. 28 gives information also concerning the relation of the peripheral pathway of taste to the trigeminus.

It must even be emphasized here, and we will be confronted by it again when studying facial paralysis, that without doubt individual variations in the course of the taste fibres occur and, indeed, it seems that their relations to the trigeminus are less regular than those comparatively constant ones to the facial nerve. The fibres emanating from the anterior two-thirds of the tongue enter almost regularly the lingual nerve, which they soon leave, however, in order to enter the facial trunk with the chorda. They leave it in the vicinity of the geniculate ganglion, mostly via the large superficial

petrosal, enter thereafter into the second trigeminal branch and with the latter reach the chief trunk. But after careful observation it must be admitted that there is a possibility of the small petrosal taking over, in a few cases, the gustatory fibres of the chorda from the facial trunk and taking them to the trigeminal trunk by means of the third branch. This covers the most important possibilities of a taste disturbance in trigeminal disease. On the other hand it would seem certain that a lesion of the trigeminal trunk need not necessarily lead to disturbances of taste. In such cases it is assumed that the fibres of the chorda after their usual course reach the

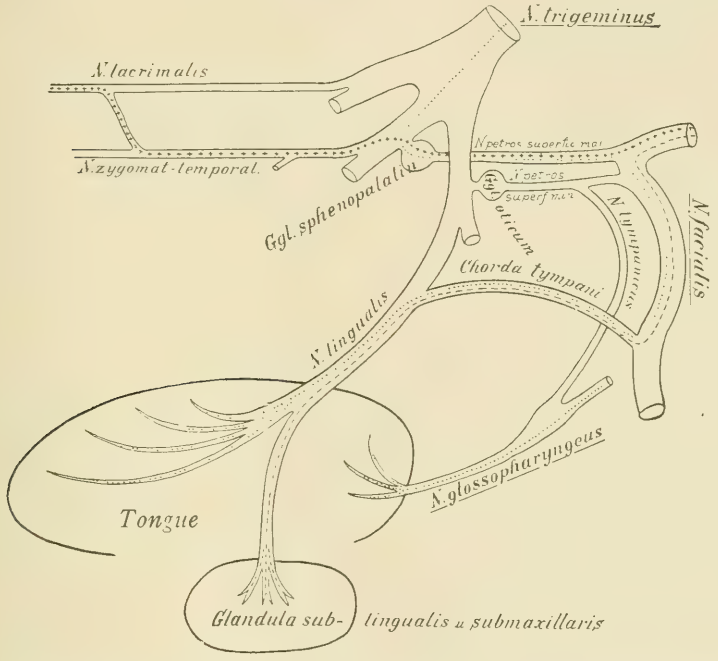


FIG. 28.—Scheme of the relations between the trigeminal, the facial and the glosso-pharyngeal. .....Taste fibres; ------Secretory fibres for sublingual and submaxillary glands; + + + + Secretory fibres for tear glands. (*Simplified from Köster.*)

glosso-pharyngeal trunk by anastomosis through the facial and with it enter the brain. That the chorda should not contain any taste fibres at all, may occur as a great rarity. In that case they either reach the trigeminal trunk directly through the lingual nerve or the lingual contains no taste fibres and the glosso-pharyngeal innervates the entire half of the tongue directly.

So far we have spoken only of the gustatory innervation of the anterior half of the tongue, usually effected through the chorda. The taste nerves of the posterior part of the tongue, which usually belong to the lingual branches of the glosso-pharyngeal, do not, perhaps, always take their further course to the brain through the glosso-pharyngeal trunk, but sometimes, by means of the Jacobson anastomosis (the tympanic, resp. the small superficial petrosal)

also enter the trigeminal trunk. This is corroborated by the experience of aurists in cases of lesion of the tympanic nerve, in connection with the hemiageusia occurring occasionally in diseases of the trigeminal trunk.

Finally, it would seem that occasionally the chorda supplies the whole half of the tongue with taste fibres.

In view of the great variability of the clinical pictures, the discussion of all these probabilities could not be omitted. The careful study of each new individual case is urged for the sake of acquiring a better and clearer insight into these complicated conditions.

Of greatest interest among the trophic disturbances are herpes zoster, to be discussed in the chapter on neuralgia, and the so-called neuroparalytic keratitis. The more or less serious inflammations in corneal anæsthesia, which are borne, however, with remarkable ease, must certainly be closely related to external injuries, from which the cornea, on account of its insensibility, is not sufficiently protected. Nevertheless the foremost authorities agree that the pathogenesis of this keratitis is still a very complicated problem. Furthermore not every corneal anæsthesia leads to keratitis.

Diagnosis.—Only the differential diagnosis needs to be discussed particularly.

The motor disturbances, when occurring isolated and without accompanying defects of sensibility, may make the diagnosis difficult.

The myopathic conditions of weakness and paralysis of the muscles of mastication—such as are found in myasthenia, in very rare cases in the common forms of muscle dystrophy, but occur more regularly in the advanced dystrophies of those affected with Thomsen's disease—manifest themselves bilaterally. The characteristic general aspects of the affections in question will always facilitate the diagnosis.

The central, supranuclear palsies of the masticatory muscles affect both sides. They are accompanied by exaggerated reflexes. R. D. is, of course, always absent. The disturbance of mastication is a partial symptom of the typical general picture of pseudo-bulbar paralysis. Therefore the diagnosis is comparatively easy in these cases also.

Difficulties in differentiation may arise in cases of nuclear and intracerebral radicular paralyses, which bear the character of peripheral paralysis, without having any features of their own, to distinguish them from diseases of the peripheral nerves. Such cases occur in focal diseases of the brain-stem, in polioencephalitis, in bulbar paralysis and in tabes. In practice, consideration of all the symptoms together will show in most of even these cases, that a peripheral nerve disease is out of the question.

The disturbances of sensation, which in their distribution correspond exactly to the innervation area of the trigeminus or one of its main branches, indicate at once a disease of the peripheral nerve. The fibres entering the pons are distributed over the long cell column of the sensory nucleus

of the trigeminal which extends down to the cervical spinal cord. If a focal lesion within the central organ affects part of this extended root area, there result disturbances in sensibility of a peculiar, segmental character; namely, stripe-like anæsthesias, which are arranged concentrically like the layers of an onion about the middle of the face. Such a zone always belongs to the areas of several peripheral nerve branches. An anæsthesia of an entire trigeminal area through intra-cerebral lesion can be thought of only in those cases in which a focus has affected the fibres already united in a bundle, immediately before their exit from the pons. It is furthermore of value to know, that in a few cases of pontile focal trouble, anæsthesias have been observed which corresponded almost exactly to those of an area of an ophthalmic nerve.

The anæsthesias in diseases of the cerebrum and in hysteria are never confined very strictly to a trigeminal area.

There can be no question of a special prognosis or therapeutics in trigeminal paralysis.

5. The Facial Nerve

The facial nerve perhaps more often than any other peripheral nerve is affected alone.

Of eminent importance in the **ætiology** of facial palsy is exposure to cold. Many authors attribute almost three-quarters of all cases to it. To be sure, patients remarkably often declare that, for instance, they looked out of the window in a heated condition, thus exposing the face to cold draughts, and that on the next day the facial palsy appeared, etc. But it is questionable, to say the least, in how many of these cases a cold is the direct cause of the disease. Recent investigations seem to favor the assumption that in not a few cases an acute, if slight, catarrh of the middle ear may be the connecting link between cause and effect.

Be that as it may, in any case the diseases of the middle and inner ear and of the petrous portion of the temporal bone are among the most frequent and most important causes of facial palsy. The thin bone, which divides the nerve canal from the middle ear, in many cases is defective in one or more places, so that the nerve is in contact with the mucous membrane of the tympanum. It is obvious from this, that even very slight disease of the tympanum, which both the patient and the physician are very apt to overlook, may lead to paralysis—a fact which in every case should prompt a careful expert examination of the ear. It must also be remembered that the palsy may continue even after the inflammation of the ear has entirely ceased. Serious and particularly purulent inflammations, and carious processes on the petrous portion of the temporal bone may injure the nerve in various ways.

In the third place, traumatic causes must be mentioned. At times, the facial is injured in an operation upon the ear, or the nerve has to be sacrificed in some indispensable radical operation. Other typical cases of traumatic facial palsy are those due to fracture of the skull, and the obstetric palsies of newly born infants, in which cases, it is usually an injury of extra-cranial branches occasioned either by pressure of the mother's pelvis, or by the forceps of the physician. The peripheral terminal arborizations of the nerve over the face suffer lesions very frequently from cuts and blows.

The last ætiological group of importance contains those palsies occasioned by basal diseases, among which meningitis of all kinds and new growths in this region play the chief rôle.

Of less frequent occurrence are lesions of the nerves in cases of adjoining inflammatory diseases located outside the cranium, such as parotitis.

In many cases facial palsy has been ascribed to some preceding infectious disease, such as diphtheria and influenza, to toxic processes, and to metabolic diseases, especially diabetes. A recent syphilis is said to predispose to facial palsy. (Tertiary syphilis causes basal palsies by meningitis.) Frequently, however, investigation does not reveal such predisposing factors. Herpes zoster of the trigeminal area may be accompanied by a facial palsy.

It is questionable, whether hereditary neuropathic affections have any significance. Some rare observations seem to support the theory of predisposition in an entire family to facial palsy. Of the relapsing forms we shall speak later.

Facial palsy appears as a congenital malformation, usually affecting both sides, frequently associated with palsy of the eye muscles and with other malformations. In the majority of cases it is doubtless due to intra-cerebral disease. The accompanying disturbances in the oculomotor apparatus may bear the character of a supranuclear or a "looking" paralysis. Like myopathic palsy, nuclear facial palsy, in which very likely must be included that occurring in cranial tetanus and tabes, is here of interest only for the purposes of differential diagnosis.

Clinical Picture

In a recent facial palsy, all mobility in the affected half of the face is more or less abolished. As a result of the flaccidity and paralysis of the musculature, the wrinkles on the affected side are smoothed out, the forehead is smooth, and the nasolabial crease not very marked. The corner of the mouth and the lower eyelid hang down a little. The mouth, as a whole, is slightly drawn toward the opposite side. The palpebral fissure is wider than on the sound side. In every expressive movement, wrinkling the brows, showing the teeth, pursing the lips, laughing, etc., only the sound side is innervated. When attempting to close the eyelids, the upper lid is lowered a trifle on

account of the flabbiness of the levator palpebræ, but it is impossible completely to close the eyelids, and the palpebral fissure remains half open (lagophthalmos). In this attempt, the upward rotation of the ball, physiologically associated with the impulse to close the lid, becomes plainly visible. In healthy individuals this can be observed only by mechanically preventing the closing of the eyelids, when the subject is strongly trying to close them (Bell's phenomenon). It may be remarked in passing, that sometimes the closing of the lids will be more complete during sleep than the patient could ever produce while awake; and, on the other hand, that sometimes the fissure is larger during sleep than during a strong volitional attempt

FIG. 29.—Paralysis of the facial at the right side during rest. (*Leipsic Medical Clinic.*)

FIG. 30.—Paralysis of the facial at the right side when trying to whistle. (*Leipsic Medical Clinic.*)

to close them. One may usually convince one's self of a unilateral palsy of the platysma by asking the patient to draw his under lip as far as possible downward. The paralysis of the posterior auricular branch coursing to the ear muscles and to the occipital muscle, and of the branches destined for the stylohyoid and the posterior belly of the digastric, manifest, as a rule, no symptoms worthy of mention.

The direct consequences of the disturbance of the mobility in unilateral facial palsy are of little importance from a practical standpoint. The patient can not whistle; when he attempts to blow out a candle, the air escapes, with but feeble pressure, through the corner of the mouth on the affected side. If there is an abundant secretion of saliva, the imperfect closing of the mouth

may become noticeably annoying, as the saliva may flow out of the mouth on the affected side.

Of much greater importance is a lasting lagophthalmos, which regularly causes the eye to weep considerably, quite often results in an obstinate conjunctivitis and once in a while even leads to graver inflammations of the eye.

In the not very rare bilateral paralysis (diplegia facialis), the motor loss is immediately and very painfully perceived. Speech is very much impaired by the loss of the labial sounds. The patient cannot expectorate and can blow only feebly. The flabbiness of the cheeks, which in chewing do not touch the teeth, makes eating difficult. The saliva cannot be properly retained in the mouth, but flows out over the lower lip. The entire face is rigid like a mask, and because of the drooping of the lower eyelids and lips, exceedingly distorted (Fig. 31).

Coincident with the symptoms of facial palsy, disturbances in secretion of perspiration frequently manifest themselves, in which case, both hyperhidrosis and anhidrosis may occur. This alone makes it evident that a strict parallel does not exist between the injury of motor fibres and those taking the same course which subserve the production of perspiration. No diagnostic interest attaches to these manifestations. The hyperhidrosis will usually be noticed when chewing, especially when chewing foods containing acids.

We now have to consider some accompanying symptoms, which manifest themselves only if the facial is injured in certain places in its course. (Cf. diagram on p. 121, Fig. 28.)

From the point where the facial nerve leaves the brain, secretory fibres subserving the lachrymal glands take their course through the nerve trunk to the geniculate ganglion, where they leave it through the large superficial petrosal nerve. A lesion of the facial nerve within the limits just outlined regularly leads to disturbance in the secretion of tears, causing either a decrease or cessation, or even a morbid increase of the secretion (Köster).

As this symptom is diagnostically important a short account of the technic for examination follows. Strips of about 20 cm. in length and 1 cm. in width are cut from blotting paper and one end of each turned down a trifle making a hook. By means of these hooks one strip is hung into the conjunctival sac of each of the two lower eyelids, which should first be somewhat dried in order to remove the too ample secretions of the mucous membrane, and their gradual permeation is then awaited. The lachrymal glands may be stimulated to action by properly inserting a small brush well up into each nostril. This test is continued, until the strips, which when fully permeated should be exchanged for fresh ones, are no longer permeated, in other words, until the lachrymal glands are "pumped out." The functional capacity of both sides is then judged according to the length of the permeated space on the strips. Minor differences (up to 3 or 4 cm. of the blotting paper) occur even in normal conditions. Now, in facial palsy we find

in the corresponding localized cases on the affected side a cessation or a large decrease, or an excessive increase, in the secretion of tears. If a marked difference between both sides is at once obvious, say if the secretion should be wholly lacking on one side, the test need not be so protracted, but in doubtful cases the "pumping out" which may take an hour or more, must be patiently awaited. Worthy of mention is the fact that the eye on the side affected with paralysis may weep, even when the secretion of the lachrymal gland has ceased. The comparatively tenacious secretion of the palpebral conjunctiva, however, is sufficient only for permeating at the most about 1 cm. of the strip of blotting paper.

The older investigators assumed that motor fibres, too, left the facial nerve by way of the large superficial petrosus, to enter the palate. Today we know that the facial does not innervate the palate. Consequently a paralysis of the palate is not a part of the clinical picture of facial paralysis. The former erroneous notion can partially be accounted for by the fact that the oblique position of the uvula, which is frequent even in healthy persons, was falsely regarded as a sign of a unilateral palsy of the palate.

FIG. 31.—Bilateral congenital paralysis of the facial. Attempt to close the eyes. Malformation of the ears. (*Leipsic Medical Clinic*).

Taste fibres which have been introduced into the facial more distally, through the chorda tympani, leave it through the large superficial petrosal, coursing to the second trigeminal branch. In an affection of the facial nerve in its course between the geniculate ganglion and the junction of the chorda, disturbances of taste are evidenced, corresponding to the innervation area of the chorda, in the anterior two-thirds of the half of the tongue on the same side.

It must be mentioned, though it does not materially affect the practical worth of the above statements, that in very rare cases taste innervation takes a somewhat different course. It occasionally happens that the chorda fibres innervate gustatorily the entire half of the tongue; but it has also been noted, that in spite of the destruction of the chorda, taste disturbances were wholly lacking.

In the area in which the chorda fibres course in the nerve trunk a very fine small branch, whose function is not known and which anastomoses with

the small superficial petrosal nerve and a motor twig, taking its course towards the stapedius muscle, are separated. It would therefore have to be assumed, that the symptoms of paralysis of this muscle exist in every facial paralysis situated above the point of separation of its twig. Indeed, increased auditory sensibility, keenness of hearing (hyperakusia), has been sometimes regarded as a symptom of stapedius palsy. But this symptom is so inconstant, so difficult to determine correctly, and so questionable in its meaning, that one has to relinquish it for all purposes of determining the diagnosis.

Fibres for the submaxillary and sublingual glands take their course down the nerve trunk to where the chorda tympani leaves. In conformity with this, in many instances, disturbances of the secretion of these glands are observed, manifesting themselves chiefly in a decrease, though sometimes in an increase in the secretion of saliva. For determining these things, we can only avail ourselves of the very inadequate method of asking the patient to raise his tongue, to dry the floor of his mouth and then to examine and compare the two sublingual carunculæ.

More pronounced trophic and vasomotor disturbances are very rare. Sensory disturbances do not, as a rule, enter into the clinical picture of facial paralysis.

According to recent investigations the chorda is supposed to contain, besides the fibres of taste, a few sensory fibres, whose further course to the central organ is unknown. It is true that occasionally a very slight deadening of the sensibility is detected in the innervation area of the chorda during facial paralysis. Perhaps slight disturbances of the sensibility in the area of the skin of the face would, with proper attention, be detected more frequently than, according to general belief, they occur. In such instances it is questionable whether it is always a true complication with trigeminal disease, or whether sensory fibres may not also enter into the facial trunk. Peripheral anastomoses between the sensory and motor facial nerves are known to exist and central relations between the facial roots and the bulbar terminals of the trigeminal fibres have been reported.

Besides disease of the chief trunk, paralyses of single branches also occur. Cuts and blows in the face are their most frequent cause. In its course through the middle ear, the chorda is sometimes affected alone as a result of an otitis media. It is occasionally destroyed in the course of an operation. As a result, corresponding disturbances of taste and of the secretion of saliva set in. Isolated paralyses of the large superficial petrosal have been observed after operative lesions in which the Gasserian ganglion has been extirpated; the scars following such procedure may also injure the nerve. We have already noted in basal syphilis the characteristic symptoms of its isolated affection—taste disturbances in the chordal area and cessation of lachrymal secretion.

The Course of the Disease

The prodromal symptoms which are sometimes observed, may be traced back in part to the original disease, such as fever, headache and earache; some of them, such as taste paræsthesia, slight clonic spasms, sometimes perhaps even pains in the face, point to a lesion of the nerve as already present.

Usually the palsy sets in acutely. A gradual onset may sometimes occur, most frequently in basal compression of the nerve.

The course of the disease, in the majority of cases, is favorable; the mobility of the palsied area is restored more or less completely. In many cases the restoration of the palsied area is not at all uniform. Some muscles are more completely and sooner able to perform their functions than others. The course of the other functional disturbances does not always correspond precisely to that of the palsy. The disturbances in the secretion of perspiration accompanying facial paralysis, are, as stated above, inconstant from the very first; they may be either paralytic or irritative and are frequently restored rather quickly. Similar conditions prevail as regards disturbances of the salivary secretion—though, it must be added, only in so far as we are able to judge. They, too, are inconstant, even when the lesion takes place in the area through which the fibres that excite the glands take their course, and they also may be restored before the paralysis. Taste disturbances are often only partial, appearing to affect only a part of the taste qualities, and they too may disappear before the palsy. One gains the impression, that possibly the taste function quite like that of sensibility in general is, in diseases of peripheral nerves, less subject to injury.

Recovery in the lighter cases is usually complete, in grave cases usually defective. The contractures and associated movements manifesting themselves in cases in which the restoration has been only partial are more troublesome than the residuum of the palsy.

The facial contracture is conditioned upon a peculiar shrinking process of the paretic muscles, thus distinguishing itself from other contractures in peripheral palsy, which are produced by the shortening of antagonists not affected by the paralysis.

The anatomic-physiological peculiarities of the muscles innervated by the facial, which to a great extent lack even a somewhat fixed point of insertion and true antagonists, may explain the exceptional position they occupy in this relation.

The contracture precisely reverses, in the later stages, the clinical picture of grave facial palsy. The wrinkles and creases on the affected half of the face, which were formerly smoothed out, become more and more clearly defined, and become even deeper than those on the healthy side of the face. The palpebral fissure becomes narrower, the corner of the mouth is lifted

up, the mouth as a whole is distorted towards the affected side. Seeing a patient in this stage of the disease, one is inclined to regard the healthy side as the paretic one. Of course, the functional test at once enlightens one.

The contracture is quite regularly attended by associated movement symptoms, which, however, also occur in the regenerative stage of cases recovering without contractures, associated movements in different muscles of the diseased half of the face, which accompany the involuntary drooping of the eyelid, and by apparently spontaneous twitchings, that pass over the face with lightning rapidity and which in reality are probably no more than associated movements due to this paralytic drooping of the eyelid, which may very easily be overlooked. Quite similar twitchings in the morbid area have been observed when bringing a hand close to the face quickly, when tapping it and when irritating electrically various places on both the sound and the affected sides: the so-called reflex twitchings. It certainly is not a matter of a genuine exaggerated reflex. The newer theory, according to which it is, in all these cases, a question of associated movement in closure of the eyelid, has many points in its favor. Also in voluntary innervation of the different parts of the facialis area, associated movements appear in other parts. In explaining all these associated movements it is very plausibly held, that in the regeneration of the degenerated nerve the fibres newly arising from every part of the nucleus, did not all find their old muscle area, but distributed themselves among various muscle areas, in consequence of which in innervations of the former, the latter also contract. This would also explain a peculiar failure in voluntary motility despite the restoration of the electric irritability, since a muscle area might have again received conductive and excitable nerve fibres, but not from its own nucleus. Therefore this area can be irritated electrically through the nerve and associated movements may also occur in this area, when innervating other parts, but every attempt at purposive appropriate innervation will fail.

The statement that in the regenerative stage of grave facial paralyses an exceedingly great confusion in the innervation takes place, very aptly expresses the actual conditions (Lipschitz).

In facial paralysis not infrequently relapses occur once or several times. The same side, or even the one which formerly remained sound, is then affected.

In deciding upon the **prognosis,** which in the majority of cases will be favorable, besides the fundamental disease the condition of the electric irritability must be carefully considered. This, though not invariably, indicates even in the second and third week with certainty whether recovery may be expected in a few weeks or only after several months. The prognosis of grave paralyses will under all circumstances be ominously affected by the associated movements, which will seldom be lacking, and by contracture.

It is often well to acquaint the patient with this prospect at any early date. The danger of a relapse is, on the other hand, of less importance.

The **diagnosis** of facial paralysis is very easy. No careful investigator will be deceived by natural differences in the two sides of the face, nor, if he note the functional disturbances properly, will he incur the danger of mistaking the side affected with paresis for the sound one, if contractures exist. Often the associated movements at once reveal the diseased side.

The real task of one who diagnoses such cases lies, first, in the distinction of peripheral nerve disease from nuclear, supranuclear and myopathic paralyses, and secondly, in determining in what part of the course of the nerve the lesion is situated.

The facial paralyses in myasthenia and in the different forms of muscle dystrophy have quite characteristic peculiarities, to which reference is made in the chapters on these diseases. As a rule they are strictly bilateral. The reaction of degeneration (R. D.) is never present. The myasthenic or dystrophic symptoms in other areas facilitate the diagnosis.

As a rule, though not always, in the supra-nuclear facial paralysis which, by the way, is usually connected with hemiplegia, the upper facial region, as a result of its bilateral cerebral innervation, remains more or less unaffected. In cases where the paralysed side takes part in a normal way in the emotionally released involuntary expression, as well as in those, where, on the other hand, the facialis is paralysed only for emotional impulses, there is clear evidence of a supranuclear seat of the lesion. Testing the reflex action in diagnosing a facial palsy, plays a subordinate part. Of all the more significance may be the electric examination for distinguishing supranuclear from peripheral paralyses. It is said that Bell's phenomenon may be absent in a central facialis paralysis. Cases are on record, in which —e. g., in diseases of the pons—a supranuclear palsy manifested itself on one side, and a palsy occasioned by lesion of the peripheral neuron, on the other. A peripheral paralysis may follow after a central one on the same side, if a focal lesion extends secondarily to the nucleus or the nerve roots.

It is sometimes very difficult or even impossible to distinguish nuclear and fascicular cases by themselves from basal-nerve diseases. The disturbances in the secretion of tears, peculiar to basal palsies, appears to be lacking in cases due to central disease. Usually, nuclear palsies are accompanied by other pathognomonic signs of disease of the central organ, or they appear in such characteristic forms, that there can be no doubt about a nuclear disease, as in the case of a bulbar paralysis, which takes a highly chronic, slowly progressing course, affecting at first, only a few muscles fibre by fibre. But even the congenital palsies, which probably depend on aplasia of the nucleus (infantile nuclear atrophy) may be confined to parts of the facialis area.

For a finer diagnosis of the place of the lesion in the area of a peripheral

nerve, the disturbances of taste and the secretion of tears are decisive. If the lachrymal secretion alone is affected, the lesion is situated basally, at all events above the geniculate ganglion. Whenever the focus is situated in the area from which the large superficial petrosal takes its origin, disturbances of lachrymal secretion and of taste will co-exist. Disease situated below the geniculate ganglion as far as the point of origin of the chorda occasions only taste disturbances. Distally from the point of origin of the chorda, disturbances both of taste and of the lachrymal secretion are lacking. In this area the paralysis frequently affects only single nerve-branches.

It should be remembered that the auditory nerve down to the geniculate ganglion takes its course in close proximity to the facial. Synchronous disease of the auditory nerve may therefore be significant for the topical diagnosis of the facial paralysis.

It is evident from the above that in inveterate cases the diagnosis will be rendered more difficult by the early disappearance of the taste disturbance. The same is true of disturbances in the lachrymal secretion.

Therapeutics.—In palsy of the facial nerve the possibility of treating the cause of the disease has to be considered first of all. In basal syphilitic meningitis, treatment by inunction promises success. In ear diseases an aurist should be consulted. In very recent "rheumatic" paralyses, some experienced physicians recommend that one or more diaphoretic procedures be instituted.

Furthermore, in the very first rank stands electro-therapeutics, applied according to the general rules for all such treatments, but especially the stabile cathodic galvanization of the nerve, and afterwards the irritant applications must be considered. Systematic exercises aided by the hand are to be recommended.

The peculiar disturbances in the regenerative stage of grave cases, as contracture and associated movements, place on the physician a difficult and peculiar task. It is an altogether erroneous impression that the contracture is caused by the electric treatment. Nor, to be frank, does it prevent its occurrence. As soon as the first symptoms appear, careful massage, with special regard to suitable manipulations for extension and flexibility should be instituted. Needless to say, this treatment should be undertaken only by a professional. To stretch the cheek and keep it extended, older doctors gave the patient a wooden ball to carry under it. The use of electricity on the sound half of the face is quite futile.

To counteract the associated movements, the patient should be taught to practise innervational exercises in front of a mirror.

The lagophthalmos of the more serious kind should be kept in mind from the start. If necessary, a protective eye bandage should be prescribed.

If an absolute paralysis is persisting in a case, and there is no longer any hope of amelioration—this will not be the case after a few months—

grafting of the nerve should be resorted to. According to the rather scanty and not particularly encouraging reports, joining the peripheral part of the facial nerve with the central end of the hypoglossus, which has been entirely divided for this purpose, can be recommended in preference to any other method. The unilateral palsy of the hypoglossus which is naturally produced thereby is less troublesome to the patient, and by a complete division, instead of a mere freshening, the vexatious associated movements between the two areas, which would otherwise set in after regeneration, are avoided. One should not hope for too much from this operation. For more detailed information cf. Lipschitz, Monatsschrift f. Psychiatrie and Neur., vol. xx, Ergaenzungsheft, page 84, and Bernhardt, Mitteil aus den Grenzgebieten, vol. xvi, page 476.

In the residual stage, plastic operations have now and then brought about a cosmetic and functional betterment. Lagophthalmos may necessitate a tarsorrhaphy.

6. The Auditory Nerve

The greater part of what we know concerning diseases of the auditory nerve, we owe to aurists. The following exposition will show how important it is that this nerve should not be neglected, as is often the case, in neurological examinations.

The auditory nerve becomes diseased either independently, primarily, or is affected secondarily in diseases of the structures that surround it.

The primary diseases are tumors, simple atrophic or degenerative-neuritic processes. Among the so-called tumors of the cerebello-pontile angle a great number emanate from the auditory nerve itself. The auditory nerve atrophies in tabes, where it is even an early symptom, and in senility (presbyacusia). Primary neuritis, comparable to neuritis optica, is observed especially in infectious and toxic processes. Among poisonings those from quinine and salicylic acid, alcohol and tobacco deserve to be mentioned. Practically more important are perhaps those cases of neuritis acustica which are caused, as has been experimentally demonstrated, by acoustic trauma or by the long-continued effect of loud noises (professional deafness of smiths, gunners). Other causes are very rare (thunderclap), or entirely problematic ("rheumatic" origin). Among the paralyses of the cranial nerves accompanying herpes zoster, a supposedly neuritic disease of the auditory nerve also occurs.

Secondary disease of the acoustic nerve we find in affections at the base of the brain, particularly in basal syphilitic meningitis, in disease of the temporal bone (caries) and especially in that of the labyrinth. Slight labyrinthine irritation accompanies various diseases of the middle and even of the outer ear. Suppurative infections of the labyrinth occur in connection with meningitis (during cerebro-spinal meningitis, after measles and scarlet

fever). Frequently they occur during infancy and later become the cause of deaf-mutism. Besides this, the labyrinth may be purulently infected from the middle ear. Traumatic injuries of the labyrinth are usually the result of indirect traumas (fracture of the skull). When caisson workers suddenly leave the place in which there is an increased air pressure, hemorrhage and gas-embolisms may result. Spontaneous hemorrhages have been observed in persons of hemorrhagic diathesis. In leucæmia, hemorrhage and leucocytic infiltrations occur. In affections of the heart the possibility of an embolus in the labyrinthine artery will have to be borne in mind. Frequently syphilis, especially late hereditary syphilis, is the cause of labyrinthine disease. Concerning the so-called choked labyrinth (corresponding to the choked disk) scarcely anything is known.

The exact situation and anatomic nature of the pathologic changes which occur sometimes in the auditory nerve after mumps are still in doubt.

Clinical Picture.—The auditory nerve is affected alone or in association with other cranial nerves. The lesion affects either both branches or only one, either the vestibular or cochlear nerve. Symptoms of irritation and of paralysis often appear simultaneously.

Diseases of the *cochlear nerve* lead to nerve deafness, the diagnostic separation of which from sound conduction deafness, occasioned by diseases of the middle ear, particularly if both forms be combined, is not always easy and frequently requires the aurist's utmost skill. The following symptoms are characteristic of disease of the auditory nerve. Complete deafness of one ear never occurs without disease of the nerve itself. It is a peculiar fact that partial disturbances of hearing affect especially the high tones, that the limit of acoustic perception is "narrowed from above," while the deepest tones may be perceived normally—an occurrence which never takes place in disturbances of sound conduction. In other cases peculiar "tone-gaps" are produced, certain parts of the tone range are lacking, which is also important in regard to differential diagnosis. The symptoms of acoustic irritation are manifold; somewhat characteristic are those taking place in the area of high tones, as the distressingly high ringing and buzzing in the ear. From tests with the tuning fork, which is held now in front of the ear, now on the skull, the following results are obtained: The duration of the perception in conduction by means of the skull is shorter when contrasted with the normal, but never, except in a preponderating complicated disease of the middle ear, lengthened (Schwabach's test). The relation between the length of perception by air and by bone conduction remains normal and the tuning fork, which is heard no longer by bone conduction, is still able to excite an auditory impulse if quickly held in front of the ear (positive effect of Rinne's test). The tuning fork applied to the crown will be heard especially in the sound ear (lateralization towards the healthy side in the otherwise rather unreliable Weber's test).

Diseases of the vestibular nerve lead particularly to disturbances of the equilibration of the body.

When one vestibular apparatus is acutely destroyed, say by hemorrhage into the labyrinth, a sudden attack of vertigo supervenes, in which the patient is prostrated, vomits, sometimes faints, usually is deafened more or less completely in one ear at the same time, in consequence of the participation of the cochlear nerve, and is tormented by violent buzzings (Menière's symptom complex). The patient has a sense of being twirled in a swing or turned round and round. He often complains about apparent movements of the objects he sees. Nystagmus towards the healthy side exists, caused or increased by directing the look that way. When, after some time, the patient tries to stand up he falls towards the affected side.

After a few days his condition is improved. The complaint usually disappears entirely after a longer time. The defect becomes latent, even a bilateral loss in the vestibular apparatus is gradually adjusted through functional compensation, and thereby becomes latent.

In chronic diseases repeated brief attacks of Menière's symptom complex may occur; the attacks are usually to be considered a symptom of irritation. The nystagmus is generally directed towards the affected side. A pronounced inclination to fall towards a certain side is rarely present. In other cases a more or less continuous feeling of dizziness is experienced, which only at times leads to a real attack of vertigo and in still others the symptoms may vary considerably. The attacks are then rudimentary and perhaps only slight, rather subjective than objective, fits of giddiness may manifest themselves.

When chronic disease has led to the gradual atrophy of the vestibular nerve apparatus, the complaints will disappear (stage of latent vestibular defect).

The **diagnosis** of disease of the vestibular nerve may, as is evident from the above, be difficult not only in the latent stage but also in some chronic cases. Though the fully developed seizure is typical, nevertheless it is sometimes extremely difficult to define the attacks and complaints in the rudimentary stage. Recently different methods for systematically testing the functional capacity of the vestibular nerve have been devised. The test for caloric nystagmus appears to be especially valuable. When syringing an ear with warm or cold water, in normal functioning of the vestibular nerve, nystagmus in the same or in the opposite direction takes place. By means of this procedure and other similar ones, it is apparently possible to diagnose both destructive and irritative conditions of the nerve with much more certainty now than heretofore. The test depends upon the absence of, or the ease with which, caloric nystagmus can be produced. For more information concerning these methods, a fuller study of which is recommended, see Barany, Physiologie und Pathologie des Bogengangapparats,

Leipsic and Vienna, 1907, and Politzer, Lehrbuch des Ohrenheilkunde, 5 edition, Stuttgart, 1908.

In *disease of the auditory nerve* the cochlear and vestibular nerves are not always affected in the same degree. The primary diseases, with the exception of tumors, frequently confine themselves particularly to the cochlear nerve and usually appear bilaterally. The majority of the secondary diseases, especially the labyrinthine affections, affect, as a rule, both the cochlear and vestibular nerves, but generally appear only unilaterally. What is known as mumps deafness, is, for the most part, unilateral and is unaccompanied by any symptoms from the vestibular area.

The **prognosis** in most forms of neuritis acustica is not unfavorable, but in almost all other conditions rather gloomy quoad restitutionem. Many diseases dependent upon acquired syphilis can be cured with specific treatment.

When disturbances of the apparatus for the perception of sound and of the vestibular nervous apparatus are found, one must consider with reference to **differential diagnosis,** besides the diseases of the peripheral nerves, also those of the intra-cerebral continuations of the auditory and vestibular pathways. An uncomplicated and complete clinical picture of an attack of Ménière's disease does indeed indicate unconditionally a peripheral affection. Otherwise, for a more minute topical diagnosis one must ask oneself, whether the accompanying symptoms point to the petrous portion of the temporal bone, to the base, the pons, or the cerebellum, as the seat of disease.

It is to be hoped that new and definite symptoms will be found in the future, by means of the new diagnostic method presented above, for the differentiation of peripheral and central lesions of the vestibular pathways.

The cochlear nerve of each side is, in the brain, immediately connected with the bilateral central auditory pathway, so that, excepting pontile focal lesions, which affect the zone of the root entrance directly, central auditory disturbances are always bilateral in origin. Experience would seem to show that only bilateral focal lesions can cause any serious auditory disturbances.

In closing, let us once more emphasize how desirable it is, that in future the disturbances caused by the eighth cranial nerve be carefully regarded even in neurological examinations. Even today they play a decisive rôle in the diagnosis of the tumors of the cerebello-pontile angle. They may be of value in an early diagnosis of tabes, and in examinations of the victims of accidents one will sometimes by this routine be enabled to decide, that besides any functional nervous consequences, organic injuries as well may have been caused by the trauma.

As to the **treatment,** the neurologist should avail himself, as far as possible, of the services of an aurist; after that he should proceed according to general principles. Especially to be mentioned is the frequent beneficial

effect of the stabile anodic galvanization in cases of sensory irritation. The button-shaped, active electrode (10 qcm.) is placed on the tragus.

7. The Glosso-pharyngeal Nerve

Concerning the diseases of the glosso-pharyngeal, but little definite is known. Isolated paralyses of this nerve have been scarcely ever observed. Up to the present time it has only been found affected in multiple basal-cranial nerve disease.

Almost invariably the lingual branches contain the taste fibres for the posterior third of the tongue. But not in every case do these fibres reach the brain in the glosso-pharyngeal trunk, for they may leave it near the petrous ganglion together with Jacobson's nerve and course to the trigeminal with which they then take their further course to the brain. On the other hand, however, it must be assumed that in some instances the taste fibres of the chorda for the anterior two-thirds of the tongue are conveyed from the facial trunk, not, as is usual, to the trigeminal, but to the glosso-pharyngeal by means of an anastomosis. Finally the possibility of the glosso-pharyngeal innervating, in exceptional cases, one whole side of the tongue can not be lightly disregarded. (Cf. Fig. 28, and the statements made concerning taste innervation in connection with the discussion of trigeminal and facial palsy.) The relation of this nerve to the function of taste is therefore undoubtedly inconstant.

For the parotid gland the glosso-pharyngeal possesses secretory fibres. They come into the gland by way of Jacobson's nerve, the otic ganglion and its anastomosis with the third trigeminal branch. Observations of aurists have revealed disturbances in the functioning of the parotid gland in lesion of Jacobson's nerve inside the tympanic cavity.

Unquestionably the glosso-pharyngeal participates in the sensory innervation of the pharynx and supplies also a part of the pharyngeal and palatal musculature with motor fibres. We have no definite knowledge with regard to the degree of this participation.

8. The Vagus (Pneumo-gastric) Nerve

Ætiology of paralysis of the vagus and its branches. Basal diseases usually injure the vagus together with its immediate neighbors, especially the spinal accessory, glosso-pharyngeal and hypo-glossal nerves. In the area of the jugular foramen, its gate of exit from inside the skull, it anastomoses with the accessory which lies closest to it. It takes up the internal branch of this nerve, and thereby receives a supply of motor fibres, which may be vagus fibres in reality since, according to several investigators, they emanate from the vagus nucleus. We have no certain knowledge regarding the function of these fibres.

Isolated affections of the vagus always indicate a lesion outside the cranium.

In the neck, gunshot wounds or those due to sharp instruments, operative injuries, lesions through pressure by glands, malignant tumors and carotid aneurisms, may all lead to palsies of the nerve trunk. But also the single branches, especially the important recurrent nerve, may become diseased in this area either uni- or bi-laterally, e. g., through tumors of the thyroid gland and new growths in the esophagus.

There are, besides, intrathoracic affections which cause paralysis of the vagus, especially of the recurrent nerve. The left recurrent winds itself around the arch of the aorta; the right, from the front backwards, around the subclavian artery. Hence the frequent injury particularly of the left nerve in aortic aneurisms will be clear. Left sided recurrent palsy, in stenosis of the mitral valve, can usually be explained by the effect of pressure of the greatly distended left auricle.

Mediastinal tumors, both primary and metastatic, pleuritic and pericarditic conditions and chronic-pneumonic affections may injure the vagus or recurrent by direct destruction, or by pressure or cicatricial contractions.

Lastly, there are cases of affection of the vagus for which no local cause can definitely be assigned. Most of these cases depend upon toxic or infectious processes. Most frequently we find palsy in the innervation area of the vagus after diphtheria, at times as a partial symptom of polyneuritis, and at times isolated. Nor can it be denied that sometimes direct injury of the nerve through the diphtheritic inflammation, perhaps a kind of ascending neuritis, occurs (Cf. diphtheritic paralysis in chapter on polyneuritis).

To what extent vagus symptoms occurring in tabes depend upon peripheral nerve disease, is doubtful.

The **symptomatology** is studied best from the point of view of disease of the single branches of the nerve which are most frequently affected alone. Symptoms of lost function are the most important and in disease of the main trunk usually stand in the foreground. Paralysis of one recurrent nerve produces motor paralysis of one vocal cord. In cases of slight lesions the paralysis is at first almost always restricted to the dilator of the glottis, the crico-arytenoideus posticus muscle (posticus paralysis). The vocal cord is adducted into a position of phonation. Inspiratory abduction is lacking, while the closure of the glottis on phonation remains unimpaired. In total paralysis the vocal cord stands half abducted, in a medial position, the so-called cadaveric-position, and is entirely immobile. The arytenoid cartilage seems shifted somewhat to the front; the vocal cord somewhat shortened and flaccid. In phonation the sound vocal cord may approach the paralyzed one across the middle line, so that a fairly normal sound can be produced. These are the two clinical pictures occurring in paralysis of the recurrent. Other paralyses of single vocal-cord muscles, such as internus paralysis,

paralysis of the interarytenoidei, are not a part of the picture of vagus paralysis, but occur chiefly in grave catarrhs of the larynx. The participation of the recurrent in the sensory innervation of the larynx is a question which has not yet been definitely decided.

Vocal-cord paralysis in disease of the vagus trunk above the point where the recurrent becomes separated, is always, as has been said, an important partial symptom of the clinical picture. If, in cases in which the seat of the lesion is higher, the fibres which leave the vagus in the superior laryngeal, enter the area of the injury, an anæsthesia of the larynx, and a paralysis of the crico-thyroid muscle, which is implicated in the tension of the cord, will result, though the loss does not produce any clinical symptoms of importance. It may be that the motions of the epiglottis depend mainly on this branch of the vagus. In very high seated paralyses of the vagus, paralysis of the soft palate and the pharynx, whose innervation is accomplished chiefly by the vagus, may be expected on the corresponding side. The sensory fibres of the pharynx also take their course with the branches of the vagus that leave the nerve trunk at about the level of the first cervical vertebra. The trouble to the patient in unilateral paralysis is comparatively slight.

The vagus also participates in the innervation of the heart. The cardiac branches partly leave the nerve trunk in the neck between the superior laryngeal and recurrent, and partly are branches of the latter. Finally, the relation of the vagus to the trachea, lungs, esophagus, stomach, liver and other abdominal organs must be mentioned.

The symptoms of the whole intestinal innervation area are, as a rule, particularly ill defined and very difficult to interpret in diseases of one nerve alone. Only when other and certain symptoms of a vagus paralysis manifest themselves, will it be possible to connect such symptoms as bradycardia, tachycardia, nausea, etc., with any degree of probability with disturbances of the vagus.

Bilateral vagus paralysis under any condition is most dangerous. Very threatening symptoms are exhibited even in bilateral posticus paralysis. Severe dyspnœa, particularly inspiratory dyspnœa, is the result of the bilateral functional loss of the abductors of the vocal cord. In grave cases, tracheotomy is necessary. In bilateral simultaneous paralysis of the superior laryngeal and recurrent nerves, the complete immobility of the larynx musculature, particularly that of the epiglottis, the inability to cough, thereby resulting, and the anæsthesia of the larynx, deprive the lungs of one of its most important protective mechanisms, and it will rarely be long before deadly aspiration pneumonia, the so-called vagus pneumonia, sets in.

Lastly, bilateral paralysis of the pharyngeal branches, whose one-sided functional loss gives rise to comparatively trifling complaints, leads to the

unfortunate condition of total paralysis of deglutition, which necessitates artificial nourishment.

Only a few words have to be added concerning *diagnosis*. A one sided recurrent, or still more, a posticus paralysis may to the inexperienced be simulated by tubercular or carcinomatous infiltrations of the larynx, that mechanically impair the mobility of the vocal cord. In the differential diagnosis of bilateral posticus paralysis, spasm of the glottis must be considered, which, however, occurs only in transitory attacks, almost exclusively in a very characteristic way in rhachitic children, and in rare instances in hysteria of adults.

A particularly important phase of the diagnosis of vagus paralysis is, as is evident from the ætiological discussion, the search for the fundamental cause of the affection.

It should be remembered that a number of diseases of the central nervous system, of which we shall name only bulbar paralysis and focal affections of the medulla oblongata, may, by destruction of the root and nucleus areas, even by bilateral destruction of supranuclear paths (pseudo-bulbar paralysis), lead to paralytic symptoms in the innervation area of the vagus.

9. The Accessory Nerve

Isolated paralyses of the accessory result only, as in the case of the vagus, through lesions outside the cranium. The lesion therefore is always distal to the anastomosis of the two nerves. The physiological significance of the anastomosis can not be estimated with certainty. In lesions common to the last cranial nerves, carious and other processes in the region of the upper cervical vertebræ must be borne in mind above all others.

In isolated accessory paralysis then, it is a matter of paralysis of the so-called external branch, which supplies the trapezius and sterno-cleido-mastoid muscles with motor fibres. The usual causes are injuries of the nerve, particularly those resulting from extirpation of tubercular glands of the neck, and inflammation and tumor formation in the immediate neighborhood.

In paralysis of the sterno-cleido-mastoid muscle, the head can be turned but feebly towards the sound side. When an attempt is made to effect this movement against resistance, it will be plainly seen that the muscle does not contract. It does not stand out sharply from the neck as in healthy persons. If a one sided paralysis exists for some time permanent shortening of the normal muscle occurs, and a caput obstipum (torticollis) usually develops.

Corresponding to the physiological function of the trapezius, which both lifts the shoulder girdle and draws it backwards at the same time, the shoulder, in total paralysis of this muscle, sinks forwards and downwards.

The medial edge of the scapula is moved outwards from the spine and runs obliquely from the inside below to the outside above (the angel wing position of the scapula). The clavicle springs abnormally forwards. Lifting (shrugging) of the shoulders, and adduction of the scapula to the spine, is possible only to a very limited extent. Through the lack of sufficient fixation of the shoulders, the functional value of the arm-movements is partially impaired. The shoulder girdle is abnormally motile on passive movement (Figs. 32 and 33).

The changes of the external form are still more strikingly evident in the atrophic stage. The neck-shoulder line, which is lengthened on the affected side, becomes angular. The acromion and clavicle, in the neighborhood of their articulation, appear to the touch to lie under the skin, as if they were part of a skeleton. The relief normal to the neck is, on the side of the atrophic sterno-cleido-mastoid muscle, flattened, even depressed.

FIG. 32.—Right-side paralysis of the accessory. Posterior view. (*Leipsic Medical Clinic.*)

FIG. 33.—Left-side paralysis of the accessory. Lateral view. (*Leipsic Medical Clinic.*)

Paralysis of the trapezius is very often incomplete, and usually some of its parts remain more or less capable of discharging their functions. Under such conditions not only should incomplete lesions of the accessory trunk be thought of, but also the fact that branches of a few spinal nerves participate in the motor innervation of the muscle and probably in an individually varying degree. On the other hand, in total paralysis, the possibility of spinal nerve branches destined for the trapezius muscle becoming paralyzed together with the accessory, must be considered. This occurs perhaps chiefly in cases in which the seat of the lesion is rather deep.

The lesion may be confined to the terminal branch destined for the trapezius, so that the sterno-cleido-mastoid remains intact.

The complication of disturbances in the innervation of the palate, pharynx and larynx, indicates intracranial disease in the region in which the accessory takes its course in close proximity to the vagus, with which it anastomoses through its internal branch. To what extent this branch participates in causing the disturbances mentioned, cannot yet, as we have before stated, be determined with certainty.

In the differential diagnosis a supranuclear paralysis will never occasion any doubt, as it invariably appears only as a partial symptom of a cerebral hemiplegia.

Nuclear paralyses are found in progressive spinal muscle atrophy and in other diseases of the spinal cord and the medulla oblongata. The rare cases occurring as symptoms or complications of tabes are probably partly nuclear, partly neuritic.

In myopathies, particularly in the various forms of muscle dystrophy, the trapezius is especially often involved; while the sterno-cleido-mastoid atrophies regularly only in advanced cases of myotonic dystrophy.

Congenital muscle defects also occur in the area of the accessory.

In considering treatment it should be borne in mind that disturbances of function may be diminished by means of orthopædic apparatus which gives support to the lax shoulder girdle.

10. The Hypoglossal Nerve

What has been said of the preceding cranial nerves, applies likewise to the hypoglossal nerve: basal intra-cranial processes and affections in the area of the upper cervical vertebræ paralyze the hypoglossal nerve together with its neighbors, while the cause of isolated paralyses must be sought outside the cranium. Injuries in the neck are more likely to lead to a paralysis of the hypoglossal nerve, but palsy now and then is occasioned by tumors or inflammation in the vicinity of the nerve trunk.

Unilateral paralysis of the tongue which is the most important result of disease of this nerve, causes the patient but slight trouble and disturbance. The tongue can be moved only imperfectly towards the sound side. As a rule it deviates towards the affected side when extended, which is usually the case when the genio-glossus muscle is involved in the paralysis—a condition which will hardly be absent in peripheral nerve diseases. When resting in the mouth, the tongue does not usually show any considerable deviation from the normal position.

On the other hand, the signs of atrophy will soon be distinctly visible, namely, the decrease in its volume, the wrinkling, the flaccid, spongy consistency of half of the tongue and further the reaction of degeneration. Remark-

ably often fascicular and fibrillary spasmodic twitchings of the diseased half and even of the whole tongue have been reported in cases of paralysis of the tongue which were surely not nuclear.

The hypoglossal nerve innervates also the so-called infra-hyoid muscles (sterno-hyoid, thyro-hyoid, sterno-thyroid, omo-hyoid). It receives the fibres for these muscles in part through several anastomoses with the superior cervical nerves. When the lesion is comparatively deep seated, those fibres, which reach the nerve only peripherally, may in part be affected also. In such cases the symptoms of the paralysis of the muscles concerned are said to be more pronounced and the hyoid bone and larynx deviate when swallowing towards the sound side; it is further claimed that a flattening of the neck in the muscle area is observable and that R. D. may be detected.

Bilateral paralysis of the tongue which, it is true, hardly ever results from peripheral causes, is of course much more serious. Eating and speaking are much hindered in such cases.

In the **differential diagnosis,** in addition to disease of the motor nerve of the tongue, supranuclear glossoplegia must be first considered. It occurs unilaterally as a partial symptom of cerebral hemiplegia and only in extremely rare cases as an isolated monoplegia. The symptoms indicative of the peripheral neuron are of course lacking. Bilateral supranuclear glossoplegias belong to the picture of pseudobulbar paralysis. Nuclear paralyses are found in bulbar paralysis and in all sorts of other diseases of the medulla oblongata. The paralysis is then very frequently a partial one; single muscles, as the important genioglossus, may remain unaffected; functional disturbances may be almost wholly wanting, and the disease may be disclosed only through circumscribed atrophy accompanied by fibrillary tremor. Tabetic hemiatrophy of the tongue is probably nuclear in some cases, neuritic in others. The radicular paralyses, too, may be partial, in explanation of which it is necessary only to consider the relatively great extension of the root-area.

Except in myasthenia, myopathic paralyses are very rare. Congenital unilateral defects have been described and also the participation of the tongue in hemiatrophia faciei.

Glossospasm, which occurs unilaterally, will not be mistaken by any careful investigator for a unilateral glossoplegia. The rigid fixation of the tongue in the spasm prevents such an error.

B. The Spinal Nerves

In the following discussion of the diseases of the spinal nerves, only those cases—selected from an abundance of possibilities—will be treated in detail which are typical and of practical importance. Those who are

familiar with the fundaments of the science of peripheral paralyses and know the innervation areas of the various nerves, will be able to judge correctly even rarer clinical pictures.

1. The Phrenic Nerve

The phrenic nerve may be injured in the neck, or in the interior of the thorax by injuries, tumors and inflammations.

The paralyses of the diaphragm resulting from the influence of toxic and infectious factors without apparent local cause, are as a rule bilateral and probably appear most frequently in polyneuritis. Diseases of this nerve are rare. For the purposes of differential diagnosis it may be well, however, to enumerate the other causes of paralysis of the diaphragm: the most varied diseases of the spinal cord in the area of the third and fourth cervical roots and the disease of these roots themselves in morbid processes of the meninges or of the vertebræ. Tabes in a few rare cases leads to diaphragmatic paralysis, which sometimes at least is of an undoubtedly spinal nuclear nature. Further there are myopathic diaphragmatic paralyses; the progressive muscular dystrophy may seize upon the diaphragm. Inflammations of the serous membrane, which covers the diaphragm, may spread to the muscle and occasion a myositic paralysis. In hysteria a picture of a complete suspension of the diaphragmatic functions may be observed. Finally, for the sake of completeness, the congenital defects which occur in the diaphragm in the peculiar form of hernia diaphragmatica should be mentioned. This latter needs no consideration in differential diagnosis.

Symptoms.—In inspiration the paralyzed diaphragm either does not descend at all or does so very incompletely. For this reason the inspiratory arching of the upper abdomen is lacking and it is even possible that during deep inspiration these regions may be drawn in. In paresis, the diaphragm is capable of a moderate though sufficient bulging; but its descent may be prevented through light pressure with the hands against the abdominal walls. Litten's diaphragm phenomenon is absent in the paralyses. The edge of the lung may be shifted somewhat upwards. An X-ray examination will show that the diaphragm stands high, that it does not bulge normally during inspiration and that, during deep inspirations, it will sometimes even be drawn upwards.

In unilateral paralysis the symptoms are well defined only on one side, while the subjective troubles and functional disturbances may be relatively trifling. Bilateral paralysis, on the other hand, must be regarded as quite a serious occurrence. Even when at rest the patient finds breathing somewhat difficult. The slightest extra demands on him occasion great anxiety. Much will depend upon the functional capacity and practice in costal breathing. A supervening disease of the lungs produces grave dangers.

The abdominal pressure is very much impaired in its influence on the viscera in the abdominal cavity through the lack of resistance from above. The abdomen, when pressed, does not arch. The force of coughing and sneezing is also diminished on account of impairment of the functional capacity of the abdominal muscles involved in paralysis of the diaphragm.

Nothing special need be added concerning the prognosis and therapeutics.

The physician should be familiar with electric irritation of the phrenic nerve because by this means in some cases of asphyxia, for instance in narcotic poisoning, the respiration and consequently the life of the patient can be kept up for several hours and time thus be gained for saving the patient.

2. The Brachial Plexus and its Nerves

The network of the arm nerves consists chiefly of the four lower cervical nerves and the first dorsal nerve. The scheme in Fig. 34 shows, how from these five roots emanate first three plexus cords and from these again, through renewed reunion and the splitting off of fibres, three secondary

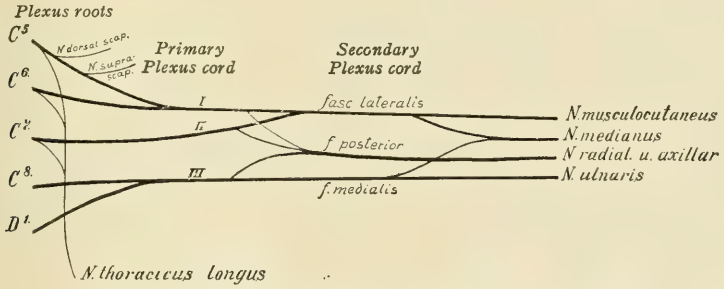


Fig. 34.—Schematic presentation of the plexus brachialis.

cords, one lateral, one medial, and one posterior. From the portion of it situated above the clavicle, the plexus furnishes quite a number of its "short" branches to the musculature of the shoulder. Below the clavicle, in the axilla, the long nerves of the arm emanate from the secondary cords, the posterior of which sends out the short axillary nerve also (Fig. 34).

As a supplement to the schematic representation, which, as far as possible, should be kept handy for reference, the following must be added. Of the short supraclavicular branches only a few have been represented. The subscapular nerve should be mentioned, which usually emanates together with several roots, from the branches of the primary bundle, that unite in the secondary posterior cord. From the medial (lower) plexus cord the medial cutaneous branches of the arm and forearm usually originate.

Although in the main the type described is regularly found, yet the structure of the plexus occasionally shows quite considerable individual variations. Thus, the fasciculus secondarius posterior, instead of arising from the primary cords, may originate from the spinal nerves themselves, of which it is constituted. The manner and place of separation of the various short branches are likewise liable to numerous variations. On that account it would be somewhat hazardous to go into minute details in a topical diagnosis of paralysis of this area.

We distinguish root paralyses, true plexus paralyses, and paralyses of the peripheral nerves in the more restricted sense of the term.

It is a case of *root paralysis* if the spinal nerves, designated above as roots of the plexus, suffer a lesion themselves, before forming the primary plexus bundle. A distinction can be made between intravertebral and extravertebral root paralyses. The most frequent causes are morbid processes in the area of the meninges or the vertebræ, traumas influencing the spinal cord directly or its immediate vicinity, such as knife cuts and localized tumor-like structures. In unilateral paralysis the symptoms of lost function will correspond to the innervation area of the affected root. This will be treated of with the spinal cord diseases.

Pluriradicular paralyses cannot, for the reasons just stated, be distinguished so easily from the corresponding plexus paralyses, unless some ætiological factors or some concomitant symptoms of the disease enable a more definite local diagnosis.

Broadly speaking three main forms may be distinguished among the *plexus paralyses:* first, the superior plexus or Duchenne-Erb paralysis; secondly, the inferior plexus or Klumpke's paralysis; and thirdly, total plexus paralysis.

Ætiology.—Mechanical injury to the plexus stands pre-eminent among all the original causes. It is produced by either direct or indirect influences.

The direct lesions usually occur in the supraclavicular fossa or the axilla. Contusions, stabs or gunshot injuries may affect the plexus; in serious trauma of the shoulder it may be affected by bone fragments or hemorrhage. The plexus may also be endangered by tumors, or even be destroyed by them; in the armpits it may be paralyzed by the constant pressure of crutches.

In indirect mechanical influences it is usually a case of traction, sometimes perhaps of a contusion occasioned by the clavicle. We shall not enter into a detailed discussion of the much disputed mechanism of the latter occurrence. It becomes particularly dangerous when the arm is drawn well down, outwards, while the head is at the same time inclined in the opposite direction. It is easy to see that in this way traction is made upon the plexus. But it is sometimes the result of lifting the arm strongly up and backwards and letting it remain some time in this position. In

both these ways plexus paralyses result, as for instance, in some exercises on a horizontal bar and in breaking in wild horses.

Dislocations of the humerus as well as brusque attempts to replace it are also significant, since especially in the latter case a direct lesion of the plexus may occur.

It is probable that in plexus paralysis of porters or longshoremen the direct pressure of the load exerts an influence besides the traction upon the plexus.

The cervical ribs are mentioned as playing a predisposing rôle.

A special position is occupied by the so-called obstetric paralysis. With infantile obstetric paralysis we have already become acquainted when discussing the facial form. Much more important and frequent are the plexus paralyses of the newly born, which occur intrapartum, and almost exclusively in deliveries by artificial means. The specific reasons are the same as in those cases of paralysis acquired in later life: direct pressure on the plexus, whether produced by fingers or instruments, more serious complicating lesions of the plexus, when a fracture of the clavicle or humerus has taken place, traction in unfortunate positions of the arms, manual traction upon an arm or awkardly releasing an uplifted arm.

Lastly, the so-called narcotic paralyses must be mentioned which may affect either single arm nerves or the plexus itself. And in these cases, too, there is no special mechanism through which they originate. The foremost cause of these unpleasant occurrences is an unsuitable position of the arms for some time during a lengthy narcosis. The feeble or non-resistance of the patient by giving rise to a favorable opportunity for a traumatic attack upon the plexus, at all events plays a greater rôle than the acute toxic process which has been assumed by some to be the principal cause of the paralysis.

Plexus paralyses founded on generally injurious, toxic-infectious influences, without any visible local cause, are extremely rare. In many cases of so-called plexus neuritis which have been observed in infectious diseases of the lungs it may be, that the main cause was a neighboring injury from an upper lobe and not a general toxic process.

By far the most frequent form of plexus paralysis is the *Duchenne-Erb "upper" plexus paralysis*. It is usually brought on by direct mechanical causes, attacking the supra-clavicular fossa and by the indirect mechanical influences enumerated before. To this group also belong most of the obstetric paralyses. The uppermost primary plexus trunk is affected. The auxiliary branch of the secondary posterior bundle emerging from the two uppermost roots is regularly affected. The deltoid, biceps, brachialis, brachio-radialis (supinator longus) muscles and frequently also the supinator (brevis)[1] muscle are paralyzed. The suprascapular nerve is very often

[1] Hence the muscles regularly affected are those that in a normal condition may be irritated together electrically from Erb's point in the supraclavicular fossa.

caught in the area of the lesion, in which case the supra- and infraspinatus are likewise paralyzed. The subscapularis is less often affected. It can be no surprise to any one who is conversant with the topographic anatomy to find that other complications may arise. But in such instances it is no longer a typical occurrence, but a more or less rare variation of the disease. The symptoms of lost function are as follows: The ability in the shoulder joint to lift the arm is partially or wholly destroyed. Unless the musculature of the forearm, particularly that flexor group emanating from the condylus internus humeri, in time vicariously assumes, as it frequently does, a part of the function, it is impossible to bend the elbow. The defect of the brachio-radialis muscle will become most obvious when the patient tries to bend the forearm, with it occupying a median position between pronation and supination, against resistance. The muscle does not project in such cases, as it does under normal conditions, from the forearm on the side of the radius, forming a characteristic edge, but on the contrary remains flat or perhaps it even sinks in, while the entire forearm seems "cylindrical." Supination of the flexed forearm (biceps) is impossible; supination of the forearm in extension becomes possible only in case of aid from the supinator brevis. Quite frequently the patient is unable, in the shoulder joint, to rotate the arm outwards (when the suprascapularis is affected). The position alone of a small child's arm may often lead to a conjecture of the nature of the disturbance; for the arm is then adducted, the elbow in extension, the forearm in pronation and perhaps the entire arm turned inwards.

Disturbances of sensibility occur in the innervation area of the fifth and sixth cervical roots which forms a broad stripe, descending on the outer side of the arm and forearm.

Klumpke's "inferior" plexus paralysis is much less frequent. It affects the plexus cord which originates through union of the eighth cervical with the first thoracic nerve. The most common causes of such paralysis are direct mechanical disturbances, tumors, and trauma. It would be difficult to state how many of such cases are, strictly speaking, plexus paralyses and how many radicular paralyses.

The paralysis may be restricted to the small muscles of the hand, the thenar and hypothenar eminences (the balls of the thumb and little finger), and the interossei. Occasionally a few muscles of the flexor group of the forearm are affected. The clinical picture becomes especially characteristic when the ramus communicans of the first thoracic nerve is drawn into the area of the disturbance. In that case paralysis of the dilator pupillæ of the side concerned results. The disturbances of sensation occupy a stripe-like area descending along the inside of the arm and along the ulnar side of the forearm. This area is innervated by the two spinal nerves mentioned above.

Total plexus paralysis affects all branches of the plexus more or less completely. As the disturbances retrogress, the paralysis may confine itself to one or another part. In some cases the area of the radial and circumflex (secondary posterior plexus cord) appears to be most seriously and lastingly injured. If the lesion has started in the axilla, the clinical picture may resemble a complete paralysis of the long branches of the arm nerves, which results, for instance, from encircling the arm too tightly.

Even when otherwise very grave and extensive, the disturbances of sensibility often leave the most proximal part of the inside of the upper arm unimpaired. This part receives sensory fibres from the intercosto-humeral nerve, which does not belong to the brachial plexus.

The diagnosis of root paralysis and of plexus paralysis in recent cases is not difficult as a rule, if a careful investigation is made and the ætiological circumstances are duly considered. Mistaking such cases for those impediments to motion resulting solely from bone lesions can always be avoided. Of course, spinal diseases situated in the primary segments of the plexus roots may lead to symptoms of lost function, which correspond more or less closely to those of the plexus paralysis. But often other signs of an intraspinal affection will manifest themselves in such cases. Serious, even insurmountable, difficulties may be found in distinguishing it from a poliomyelitis that is restricted to an arm area, though probably only in such inveterate cases in which no information can be secured, concerning the conditions under which the disease developed. The absence of disturbances of sensation, which latter almost without exception in poliomyelitis remains normal from the very beginning, does not afford a differentiating criterion in inveterate cases. Judging residual conditions is made particularly difficult by the fact that after partial recovery the defects may confine themselves irregularly to some single muscles.

It must not be forgotten, besides, that both intra- and antepartum cerebral and spinal diseases and paralyses of single peripheral nerves may be acquired in some rare cases. Not every paralysis affecting the arm of a newly born child is a plexus paralysis.

Statistically considered the **prognosis** of a plexus paralysis is decidedly less favorable than that of a single peripheral nerve paralysis. Perhaps this is due to the fact that the anatomic injuries are much more serious than is usually supposed. For this reason an early, careful and active **treatment,** and a vigilant observance of the course of the disease are urgently demanded in every case. Especially in these cases is any negligence, and particularly a "laisser aller" particularly reprehensible. It is self evident that complicating bone lesions require surgical treatment. Even the nerve injury itself may necessitate an operation. If there was no occasion in the early stages for suturing the nerve, nevertheless, if the neurological treatment remains without results for some time, one should consider the feasibility of finding

the plexus secondarily, and if needed, paving the way for the restoration of function by excision of scars and by suture. The results obtained by such treatment have been encouraging.

It is well to state, that especially on account of the peculiar irritation conditions obtaining in the first few weeks after birth, electric prognosis in affections of the newly born does not as a rule give reliable results.

A knowledge of the origin of plexus paralyses should induce the surgeon and the obstetrician to use whatever prophylactic care may be necessary.

The Single Peripheral Nerves That Emerge from the Brachial Plexus

a. Short Branches Originating Above the Clavicle

The only paralysis in this group of any practical significance is that of the long thoracic nerve, because it alone appears more frequently independently. Its causes are similar to those of the plexus paralysis: mechanical influences in the supraclavicular region and in the axilla, certain large, straining, long continued movements, in connection with traction upon the arm, especially raising the shoulder joint. In some cases generally injurious, toxic infectious influences occupy a prominent place.

The clinical symptom of this paralysis is the paralysis of the large serratus anticus muscle. As a result of the preponderant pull of its antagonist, the shoulder blade, when at rest, usually stands somewhat higher and its inner edge is nearer to the spinal column than on the sound side. The inferior angle of the shoulder blade usually stands off a little from the thorax, and thereby slips out from the covering formed by the edge of the latissimus dorsi. The ability to raise the arm sideways and especially forwards is seriously impaired; in fact, the arm as a rule cannot be lifted higher than a horizontal position. In time, however, a far reaching compensation may take place, in which numerous muscles, among which the supraspinatus and trapezius muscles stand first, participate. In all cases, however, the serratus does not contract when the arm is raised, which is revealed by inspection and palpation of its indentations. The characteristic turning of the shoulder blade which moves its inferior angle outwards and upwards, and which, occasioned by the serratus, sets in as a rule not when the arm is beginning to be lifted, but shortly before it reaches a horizontal position, is likewise lacking. Instead of this, the above mentioned anomalies in position, which even when at rest are usually visible, become more prominent when the arm is raised sideways; in particular the medial edge of the scapula moves close to the spinal column. In addition to this, in raising the arm forwards, the medial edge stands off considerably backwards from the thorax, so that it is possible to feel the under surface of the scapula or the subscapular muscle. This position has been designated as the "winged" position of the scapula (Fig. 35).

In diagnosing the paralysis, particular dependence must be placed upon the absence of visible and palpable contractions of the muscle and also upon the absence of the characteristic movement of the scapula when it is attempted to raise the arm. An over-estimation of the anomalies in position has led to the adoption of a pseudo-serratus paralysis. In some very thin persons a kind of "wing" position of the scapula may be observed, which in asymmetrically built people may appear even unilaterally, when no serratus paralysis exists. In such cases it is very easy to prove that the muscle is capable of performing its functions.

FIG. 35.—Right-side paralysis of the serratus. Elevation of both arms laterally and slightly forwards. (*After Curschmann-Schüffner, Leipsic Medical Clinic.*)

For persons engaged in manual labor a serratus paralysis is a serious defect, but the prognosis, in which one should consider the possibility of compensation, is not on the whole unfavorable.

The *other supraclavicular branches* of the plexus are, indeed, seen in plexus paralyses some more and some less often affected, but their isolated paralyses are neurological rarities, which are observed now and then especially after mechanical injuries of the primary area of these nerves. Defects in the area of the muscles innervated by them are usually caused not by a disease of the nerves, but by myopathic-dystrophic, and above all, by arthritic processes (arthritic atrophy of the supra- and infraspinatus), and central

affections. Congenital defects, particularly in the pectoral area, may also occur. In the latter cases the compensation is often remarkable for its completeness.

b. Branches Developing From the Infraclavicular Plexus Section

Circumflex Paralysis.—Ætiological factors are traumatic influences in the shoulder and supraclavicular areas, pressure acting in the axilla, traction upon the nerve occasioned by excessive raising of the arm and by dislocation of the shoulder-joint, and in some cases by noxious factors of a toxic and infectious nature. In those cases in which the paralysis results from sleeping upon the uplifted arm, as well as in infantile obstetric paralysis and that produced during narcosis, pulling or stretching the nerve is the chief cause.

The clinical symptoms are paralysis of the deltoid muscle and anæsthesia in the cutaneous area supplied by the nerve. The muscle cannot be made to contract; neither can the arm be raised from the trunk. The occasional preservation from injury of an anterior muscle segment which derives its fibres from the anterior thoracic nerve, is of no special functional value. On the other hand, in the later stage, a partial functional compensation may be possible by means of numerous auxiliary muscles, among which the supraspinatus, which is specially important in the earlier stages of the movements, stands foremost.

The paralysis of the teres minor, which is likewise supplied by the circumflex, cannot, as a rule, be shown.

In regard to differential diagnosis, the fact is very important that the deltoid muscle exceedingly often undergoes an arthritic atrophy, while its nerve remains unimpaired. To determine such cases, the affection of the joint should be proved first of all. In arthrogenous atrophy, both the supra- and the infraspinatus are regularly affected, R. D. is absent, and, of course, the disturbances of sensation in the circumflex area are always lacking.

Further, direct myopathic paralyses of the deltoid muscle resulting from blows or pressure upon the muscle (as from lying upon the shoulder) have been described, and these, too, might lead to a confusion between the two.

Because of its rarity, we shall do no more than simply mention the *paralysis* of the *musculo-cutaneous*.

Musculo-spiral or radial paralysis is among the most frequent peripheral paralyses. It owes its special predilection to the extremely exposed situation of the nerve. During the greater part of its course along the arm, the musculo-spinal nerve lies directly on the periosteum of the humerus, and particularly where it entwines itself around the bone on the external side of the arm is it exposed to all kinds of injuries. In the first place, bone fractures in the most diverse places may affect the nerve at once, as well as lead later to so-called secondary traumatic paralysis. Further, the nerve may

suffer lesions from pressure in its course along the arm, in the armpit from crutches, and in different places through an unfortunate position of the arm during sleep or narcosis. Sometimes stretching plays a part, as when the arm has lain in an extremely raised position. Dislocation of the shoulder joint may also cause a radial paralysis. It is superfluous to enumerate the many possibilities of direct lesion by blows, shots, etc. Paralysis by constricting cords through encircling the upper arm, which affects the radial in particular, has already been mentioned. Now and then, paralyses are occasioned by bungling injections of ether or other substances, or by injections in unsuitable places. In some cases, the paralysis has been due to a sudden violent contraction of the triceps muscle.

In individual predispositions, such as exist, e. g., in toxic conditions and in tabetic individuals, frequently very slight, external local influences suffice to occasion the paralysis.

The so-called lead paralysis of the musculo-spiral nerve will be discussed with polyneuritis.

Clinical Picture.—In the vast majority of cases, the paralysis affects only the group of extensors of the forearm, and the disturbance of sensation is limited to the skin area of the hand, which is supplied by the musculo-spiral nerve. The motor symptoms of lost function are as follows: Extension of the wrist and of the fingers—except where rendered possible by means of the interossei and lumbricales muscles—the supination of the extended arm and abduction of the thumb, in so far as not enabled by the abductor brevis, become impossible. An important synergist for bending the elbow was lost in the brachio-radial muscle (supinator longus), whose defect can be ascertained best, if, as has already been mentioned when discussing Erb's paralysis, the patient is made to flex the forearm, when in a semipronated position, forcefully against resistance. In a normal condition, the muscle on the radial side of the forearm will then project sharply, while in a radial paralysis no contraction whatever occurs.

This loss, however, impairs also the functional capability of a number of nonparalyzed muscles. Doubling the fingers into a fist can be accomplished but feebly, if the synchronous extension of the wrist be wanting. The ability of the patient to spread his fingers out normally, and to execute lateral movements of the wrist, can be demonstrated only when his fingers and hand have been assisted to retain that semi-extended position favorable to this function, but which they cannot assume by themselves.

When the hand is left to itself, it hangs down limp from the wrist, the fingers are somewhat flexed and the thumb is slightly opposed (wrist drop).

Only comparatively rarely, when the seat of the lesion is very high, does the triceps muscle share in the paralysis. In such cases the disturbance of sensation may also affect the cutaneous innervation area of the musculo-spiral on the forearm (N. cutan. antibrach. dorsalis) and even on the arm

(N. cutan. brach. posterior). The participation of the musculo-spiral nerve in the innervation of the brachialis anticus muscle (outer part) may under certain circumstances be shown conclusively by electric examination.

That the motor paralysis from the beginning is restricted to part of the muscles of the extensor group, is likewise observed only comparatively seldom. This form is most likely to occur when the lesion is situated on the forearm.

The disturbances of sensation in musculo-spiral paralysis are usually trifling.

A peculiar form of trophic disturbance, manifesting itself as a thickening of the tendons and which is designated as Gubler's tendon swelling, has been observed particularly in musculo-spiral paralysis, in the region of the back of the hand. It does not seem to be of any great importance.

The **prognosis** of musculo-spiral paralysis, statistically considered, is exceedingly favorable. Because of the exposed position of the nerve, a particularly large number of cases of slight paralysis are observed. It has been shown convincingly, that especially in musculo-spiral paralysis, early and proper electric treatment affects very favorably the prognosis in regard to the time required for recovery (E. Remak).

As a supplement to the general rule for the **treatment** we add that in incurable musculo-spiral paralysis, orthopedics has at its disposal, besides operative treatment, suitable corrective apparatus constructed as cuffs with attached gloves, serving to replace by rubber traction the loss of the extensors, which is so troublesome in using the hand.

Median Paralysis.—The various kinds of mechanical injuries which, as we have seen, threaten the musculo-spiral especially in the area of the arm, may all (with the exception of a paralysis caused by sudden muscular contraction) affect the median also, though on account of the protected position of the nerve this occurs much less frequently. Among the most frequent causes of median paralysis is complete division of the nerve by wounds resulting from cuts or stabs on the anterior side of the forearm. Ascending neuritis in consequence of infected wounds in the area of the terminal ramifications has been observed particularly in the median.

If the nerve sustains a lesion in its course down to the elbow joint, there ensues a paralysis of the entire flexor group in the forearm with the exception of the muscles supplied by the ulnar, of the flexor carpi ulnaris, and of that part of the flexor profundus digitorum destined for the two last fingers; further, a paralysis of the ball of the thumb (excepting the adductor pollicis muscle) and a disturbance of sensation in the greater part of the palm of the hand belonging to the median nerve.

The alteration of motor functions is therefore as follows: Pronation of the forearm becomes impossible. The wrist can be bent feebly only when aided by a simultaneous abduction toward the ulnar side. Flexing the second and

third phalanges is possible only in the last two fingers and even there only to a limited extent. By means of the interossei and lumbricales, flexing the proximal phalanges from the second to the fourth finger, under synchronous extension of the other joints, remains intact. The apposition of the thumb disappears.

Especially as a result of this disappearance the hand assumes that peculiar position, which has caused it to be designated as ape-hand.

Generally speaking the disturbances of sensation are much more strongly pronounced in median paralysis than in other peripheral paralyses of the extremities.

If the median suffers an injury not far from the wrist, which happens not infrequently, the clinical symptoms are restricted to the disturbance of sensation and to the paralysis of the ball of the thumb—still, however, with the exception of the adductor.

Ulnar Paralysis.—What has been said concerning the mechanical ætiology of median paralysis is true also of the ulnar. It must be added, however, that besides being exposed to mechanical injuries on the flexor side of the forearm, where it is not seldom injured, e. g., by cuts, the ulnar is especially exposed to them also in the region of the internal condyle of the humerus. Above all pressure paralyses occasionally result from lesions at this point, for instance when during sleep the head is propped up by the elbow. A particular predisposing condition for this arises, when the nerve is not securely seated in a deep sulcus, but, being in a position of habitual dislocation, easily slips from its resting place. This condition appears either congenitally or, in rare cases, is due to traumatic influences. Paralyses of the ulnar, apparently originating spontaneously, especially in persons predisposed to them through toxic influences or infectious processes are described likewise (Fig. 36, *A* and *B*).

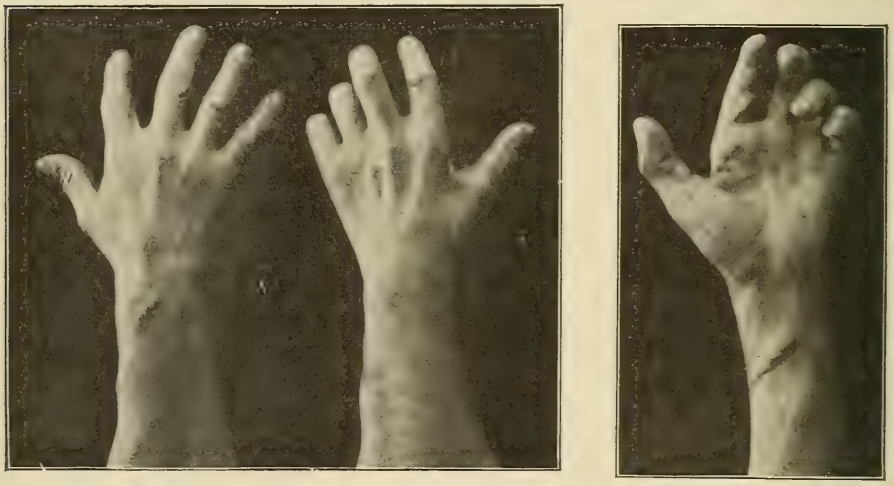
In injuries located in the lower part of the forearm, the paralysis is restricted to the interossei and lumbricales, the muscles of the ball of the little finger and to the adductor pollicis. The pure adduction of the thumb, both the abduction and adduction movements of the other fingers, and, moreover particularly the flexion of the proximal phalanges of the second to the fifth fingers with synchronous extension of their second and third joints are suspended. The last named function is usually impaired less in the second and third fingers than in the other two, as the first two of the lumbricales, which just in this movement participate as synergists of the interossei, as a rule, derive their motor fibres from the median.

In high seated lesions of the ulnar nerve, the flexor carpi ulnaris, which bends the wrist towards the ulnar aspect of the arm, and the flexor profundus digitorum for the last two fingers, which participates in the flexion of their second and third phalanges, are also paralyzed. The sinking in of the interosseous spaces and the atrophy of the ball of the little finger are

accompanied then by a characteristic flattening of the ulnar aspect of the forearm.

The typical position of the fingers, overextension of the first phalanges and flexion of the others from the second to the fifth fingers, dependent upon the preponderance of healthy antagonists, is designated as claw or talon hand, main en griffe. The anomaly is most strongly pronounced in the fifth finger, least in the second, as can be easily understood from the above statements.

The disturbances of sensation occur in a greater or smaller part of the innervation area of the ulnar nerve situated on the ulnar side of the hand. The ulnar edge of the hand and the small finger are rather regularly hypæsthetic.



A *B*

FIG. 36.—*A*, paralysis of the left ulnaris, caused by the division of the nerve in the forearm. The normal hand alongside. *B*, the affected hand from the volar side. Atrophy of the hand-muscles supplied by the ulnaris, claw or talon hand. The formation of the usually present hyperextension of the basal phalanges of the ring and little fingers was prevented in this case by traction from the scar located at the flexor side of the forearm. (*Leipsic Medical Clinic.*)

Ulnar paralysis seems to cause a certain predisposition to Dupuytren's fascia contraction.

When, in the general part, the reader was cautioned against confusing a peripheral paralysis with all kinds of malformations and impairments of movements, mechanically produced by cicatricial, arthritic, and other processes, which may be accompanied by muscle atrophy and even slight numbness of sensation, it was really more particularly the ulnar paralysis that was alluded to. Careful examinations will prevent such errors.

In the **treatment** of ulnar paralysis, the habitual dislocation of the nerve must be considered in some cases. It may be remedied by various operative methods.

3. The Other Thoracic Nerves

It is extremely rare that paralyses of this area come under clinical observation. Now and then paralyses of the abdominal muscles result from a disease of the lower thoracic nerves, chiefly occasioned by pathological processes that injure the roots concerned in the area of the spinal meninges. Sometimes, however, they may appear in connection with infectious factors and herpes zoster.

Motor disturbance is most clearly shown in the attempt to exert abdominal pressure. The entire abdomen, or the diseased side, is spherically arched, but no contraction of the abdominal muscles can be seen or felt. It becomes impossible to raise the trunk from a lying position without assistance from the hands. In this attempt, too, the inability to exert the abdominal muscles can be clearly seen and felt. In unilateral paralysis the umbilicus, which even when at rest sometimes deviates somewhat, is drawn considerably towards the sound side. The abdominal reflex is absent in the area of the disease, and hypæsthesia may usually be shown to exist.

Complete paralysis of the abdominal muscles leads to the development of an abnormally increased lordosis of the lumbar portion of the spine. More serious is the impossibility of making strong expiratory movements.

Much more frequent than the peripheral paralysis of abdominal muscles is their participation in progressive muscle dystrophy and in varied spinal diseases. Congenital defects also occur.

4. Nerves of the Lumbar and Sacral Plexuses

The plexus of the leg (plexus lumbo-sacralis) arises from the lumbar nerves and from the first, second and half of the third sacral nerve. These roots of the plexus participate in the formation of the cauda equina, and proceed by a more or less extensive course along the inside of the spinal canal, where diverse pathological processes may injure them. Radicular paralysis of this area resulting from such injury will be dealt with in the chapter on spinal diseases. After their exit from the intervertebral foramina, the first three lumbar nerves and half of the fourth unite to form the trunk of the crural nerve, after they have sent out a number of secondary branches: a few small ones, mostly sensory, and the more important obturator and lateral femoral cutaneous nerves (external cutaneous). So much for the area of the lumbar plexus. Now, the other half of the fourth lumbar nerve, having united with the fifth to form the lumbo-sacral cord, coalesces with the sacral nerve segments of the leg plexus into the sciatic nerve. The roots of this sacral plexus area send out several secondary branches, namely, the gluteal nerves, the posterior femoral cutaneous (perforating cutaneous) and the motor nerves for the small outward rotators of the thigh, the piriformis, obturator internus, two gemelli and the quadratus femoris muscles. The

branch for the first mentioned rotator muscle always emanates from the roots; the others may arise from the sciatic trunk (Fig. 37).

Concerning peculiar groupings of symptoms, which would correspond to lesions of certain plexus segments and which might be clearly differentiated from pictures of paralyses of peripheral branches, we do not possess the definite knowledge that has been obtained for the brachial plexus. In affections of the leg plexus, we are usually confronted with the same clinical pictures as in cases of single nerve paralysis, such as crural paralysis, sciatic paralysis, etc. Some few remarks, which will perhaps be of value for the localization in the plexus area, will be given in the following paragraphs dealing with paralyses of the more important single nerves.

Anterior Crural Paralysis.—Among the more important causes of this rare paralysis are the following: Injury of the nerve by direct wounding, through tumors, particularly those near the spinal column, in the pelvis, also by means of the pressure which the nerve may sustain from the body of the child during protracted delivery (maternal obstetric paralysis); pressure injuries caused by protracted extreme flexion of the hips, such as have been observed in infantile obstetric paralysis in children born with breech presentation, or after narcoses during which the leg was awkwardly placed; lesions of the nerve in fracture of the pelvis and in fractures and dislocations of the thigh. Lastly, a crural paralysis may in some cases be observed without any visible local cause in persons who were under the influence of metabolic disturbances or of infectious and toxic processes.

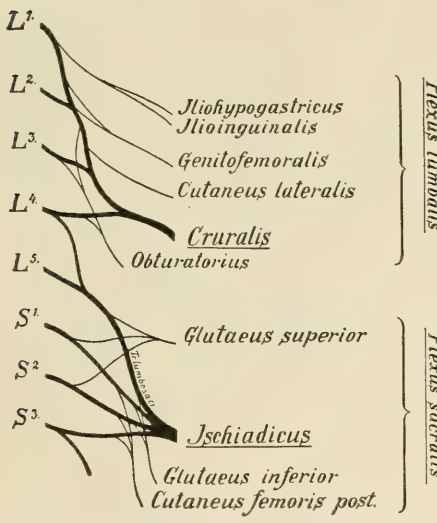


FIG. 37.—Scheme of the lumbo-sacral plexus.

The most important and the most regularly occurring consequence of a crural paralysis is the paralysis of the quadriceps femoris muscle. Active extension of the knee joint becomes an impossibility. The patient can support himself on the leg only while he keeps the knee in a passively forced extended position. As soon as it is flexed, he collapses. For this reason he swings the diseased leg forwards when walking, and after he has put down the foot, passively presses the knee into extension until the other leg again supports the body. The paralysis of the sartorius can be determined by inspection and palpation; it does not, however, occasion any considerable functional disturbances. The paralysis of the little branch, through which the anterior crural participates in the innervation of the pectineus, appears

to have no significance. In high seated lesions the ilio-psoas is likewise paralyzed. This results in a loss of flexion in the hip and walking is rendered extremely difficult. A bilateral quadriceps paralysis also renders walking very uncertain, while in a bilateral, complete paralysis of the anterior crural nerve walking becomes altogether impossible.

Besides motor disturbances, sensory ones in the extended area of the skin supplied by the anterior crural nerve are part of the disease picture. The patellar tendon reflex is destroyed.

It is hardly necessary to emphasize that a concomitant affection of the secondary branches of the lumbar plexus may sometimes acquire a topical-diagnostic significance. For parts of the innervation area of this peripheral nerve to participate in the most diversified and different, myopathic and nerve diseases occurs much more frequently than its rare paralysis. One of the most frequent causes of a weakness resembling paralysis in the quadriceps area, is an arthrogenous atrophy of the muscle in diseases of the knee joint.

Therapy.—It must be emphasized that an orthopædic apparatus, which replaces the functions of the lost muscles by elastic bands, may materially improve the ability to use the leg.

Obturator paralysis is still more uncommon. It manifests itself alone or accompanies anterior crural paralysis. Ætiologically affections in the pelvic area must be mentioned. An obturator hernia may lead to paralysis of the nerve; both maternal obstetric and narcosis paralyses have been observed.

The adductors of the thigh, of which only the pectineus and magnus are slightly innervated from other sources, are paralyzed, and so is the obturator externus, which participates in the external rotation of the thigh. Besides this, disturbances of sensation may manifest themselves on the inside of the thigh.

The **paralysis of the lateral femoral cutaneous nerve** (external cutaneous) is of greater practical interest. There is a peculiar clinical picture, known as *meralgia paræsthetica*, Bernhardt-Roth's disturbance of sensibility, caused by an isolated destructive disease of the lateral femoral cutaneous nerve. Usually it appears unilaterally; patients complain of various unpleasant sensations and pains on the outer side of the thigh, which become particularly pronounced when walking. All mechanical irritations, even the pressure of the clothes, become very unpleasant. Some patients even complain of a feeling of heaviness in the calf, manifesting itself when they walk and somewhat resembling intermittent claudication. Objectively, a hypæsthesia or anæsthesia exists in a smaller or greater part of the innervation area of the nerve. The seat and extension of the anæsthesia may fluctuate. Very often pressure will reveal particularly painful points along the course of the nerve.

The ætiology of the disease is obscure. It develops in connection with infectious diseases, such as typhoid and articular rheumatism. It has been seen to follow injurious mechanical influences and overexertion, and in patients suffering from disordered metabolism, and it has been ascribed, more or less correctly, to several other factors. The affection is of a harmless nature; nevertheless it may become very troublesome to quite a few of those who suffer from it. The prognosis is very questionable, for the disease may be very obstinate and even relapses may occur.

Of therapeutic measures, faradization may often be recommended, though in cases with great irritation the stabile anode galvanization, in which the inactive electrode can be placed on the calf, is usually to be preferred. It has been reported that in some cases a resection of the nerve was successful.

Sciatic Paralysis. Ætiology.—Even in their course through the pelvis the sciatic fibres may sustain various injuries. One of the most important causes is maternal obstetric paralysis of the nerve, occasioned by the pressure to which in protracted delivery the nerve is exposed. Besides lesions may result from tumors, inflammatory processes of neighboring organs, or pelvic exudates from fracture of the pelvis and of the lowest part of the spinal column. Traction and stretching are produced especially by dislocations of the hip; sometimes also by careless attempts to reduce them. At the time when brusque stretchings of the nerve were still used as a treatment of neuralgia, paralyses were seen to result from them as well as from remedial injections into and below the gluteal musculature.

Further, peripherally the nerve is especially endangered by fractures of the femur.

In the upper part of the popliteal space the two terminal branches, the peroneus and tibialis nerves (external and internal popliteal), separate. Along their individual course they are exposed to numerous dangers, which require a special discussion.

The peroneal nerve (external popliteal nerve) is affected especially, frequently by pressure influences, as for instance, in occupations which must be carried on in a stooping, squatting position (e. g., workers in sugar beet fields), by pressure during narcosis or sleep and in particularly predisposed persons, even from keeping one leg over the other for a long time. Traction, for instance from a sudden twist in the ankle, affects in particular the superficial branch. Frequently fractures of the leg, especially of the fibula, affect the nerve; but on the other hand, affections of the knee joint, in spite of their proximity, only extremely seldom exert a harmful influence. The equally rare case of an infantile obstetric paralysis of the peroneus nerve occasioned by delivery of the child by the foot is mentioned on account of its prophylactic interest.

The tibialis nerve (internal popliteal nerve) is much less endangered. In its more protected position it suffers less frequently from pressure lesions;

probably the most frequent cause of its isolated paralysis is a direct trauma.

The nerves of the sciatic area are also sometimes found to be affected with an isolated paralysis, though no local cause can be determined. As a rule, a toxic or infectious condition or a metabolic disturbance may be shown to have been a strongly predisposing factor. Connected with protracted neuralgia of the sciatic nerve are often symptoms of lost function, and even more or less pronounced paralysis symptoms.

The discussion of **symptomatology** begins most appropriately with a discussion of the symptoms of peroneal and tibial paralysis.

(a) **Peroneal Paralysis.**—In complete paralysis the extension (dorsal flexion) of the ankle and of the proximal phalanges of the toes as well as the pronation (abduction) of the foot are entirely lost. Extension of the second and third phalanges is considerably impaired and is now possible only with the aid of the interossei and lumbricales, which at the same time flex the proximal joints. Supination (adduction) of the foot is seriously impaired by the loss of the synchronous extension of the anterior tibial. The foot assumes a pes equinus position, while the toes are flexed. To prevent the toes from dragging when he walks, the patient has to lift his knee to an abnormal extent (steppage gait[1]). In protracted paralysis and with insufficient medical aid, the abnormal position of the foot will become permanently fixed by the contracture of antagonists. Since the pronation of the foot is completely suspended and yet a few supinators are preserved (those supplied by the tibial nerve), there is added to the pes equinus position a slight raising of the medial border of the foot. Consequently a pes equinus varus is formed.

Disturbance of sensation affects the dorsal surface of the foot and toes and a short adjoining portion outside on the extensor surface of the leg. The cutan. suræ lateralis nerve (communicans peronei) which originates in the popliteal space immediately after the division of the peroneal and the tibial nerves, is not considered in this discussion of isolated peroneal paralysis.

It has already been mentioned, and it will be enlarged upon when discussing sciatic paralysis, that the two main branches of the peroneal nerve do not always participate to the same extent. Even direct injuries may affect only one of them. The superficial branch (musculo-cutaneous) innervates only the peronei muscles and but a very small part of the region of the skin. Everything else devolves upon the deeper branch (anterior tibial nerve). To understand the functional losses and anomalies of position manifesting themselves in cases of partial paralyses of the peroneus region, it must be remembered that in paralysis of the anterior tibial the foot is disposed to assume a valgus position; in paralysis of the peronei

[1] Charcot compared this gait to that of the grave step of noble horses; steppeur, Engl. stepper, from to step, meaning particularly a dignified step.

muscles, on the other hand, it tends to assume a varus position. The peroneus longus muscle which is inserted at the base of the first metatarsal bone, is also an antagonist of the tibialis anterior muscle in so far as it lowers its point of insertion anteriorly and increases the arching of the foot. If it is paralyzed, a flattening sets in; in case of its contracture an increase of the arching of the ankle. The function of extension devolves solely upon the ramus profundus.

(b) The **tibial paralysis** affects the musculature of the calf, the muscles of the balls of the big and the little toe, the interossei and lumbricales. Flexion of the foot and toes is lost—the slight flexor action of the peroneus longus is hardly of any value in view of the grave defect—the patient is unable to spread out his toes or to adduct them; neither can he extend the second and the third toe joints with a synchronous flexion of the proximal phalanges. Supination is possible only by means of the tibialis anterior, therefore only with simultaneous extension. The foot assumes slight extension and pronation (i. e., calcaneo-valgus) position; the toes in the metatarso-phalangeal joint suffer over-extension (claw foot, pied en griffe).

The disturbance of sensation occurs in the plantar region of the foot, including the flexor surface of the toes and the outer area of the ankle, perhaps even a short contiguous portion outside on the posterior aspect of the leg.

The Achilles tendon reflex is destroyed. The plantar reflex is either destroyed or at any rate, if the sensory conduction is preserved, abnormal. The characteristic flexion of the toes is suspended on account of an interruption in the motor path. The extension of the toes which takes its place, especially in the case of the big toes, does not, of course, have the diagnostic significance of the Babinski reflex (pseudo-Babinski).

(c) **Sciatic Paralysis.**—If the nerve suffers a complete interruption of conductivity above the place where the separation of the peroneal and the tibial takes place, it will result in an absolute paralysis of all the muscles in the leg proper and in the foot. In high seated lesions, the flexor muscles of the thigh (biceps, semimembranosus and semitendinosus) are also paralyzed. The loss of the little branch by which the sciatic participates in the innervation of the adductor magnus muscle, and the paralysis of the small outer rotators, will hardly occasion any considerable clinical symptoms.

The disturbance of sensation extends also into the area of the cutaneus suræ lateralis nerve (external saphenous nerve).

Especially important is the fact that in high seated lesions in the sciatic area, partial paralyses, particularly in the area of the peroneus, occur. It is almost always the case in maternal obstetric paralysis, that any existing more extended defects soon retrogress, but that a peroneal paralysis, which shows a decided preference for the area of the ramus profundus, thus sparing the peronei muscles, remains. A great many attempts have been made to

explain this peculiar condition. The cause for the last mentioned specific case might be traced to a certain part of the plexus (which was mentioned in the introduction as the lumbosacral cord) which passes into the sciatic and which perhaps carries fibres for the ramus profundus peronei, that is especially exposed to mechanical injuries in the pelvis. At any rate, we are really confronted with a greater liability to lesion in the peroneal than in the tibial nerve. Whether it is exposed more to mechanical injuries in other places than in the one named, whether more unfavorable vascularisation conditions obtain, or whether still other factors have any part in it, is unsettled. An anatomic fact, which may some time acquire clinical interest, is the following: even though the fibres destined for the peroneus and tibialis always take their course as far as the upper part of the popliteal space in closest proximity, yet they are frequently to be found far up from the point of their separation, and even as far back as the plexus area, as separate, or easily separated bundles.

In the **prognosis,** one should be very careful not to interpret too optimistically those sciatic paralyses, whose point of lesion must be looked for in the deeper part of the pelvis. Particularly the maternal obstetric paralysis leaves, in a great percentage of cases, a lasting defect.

In separating numerous peripheral paralyses in the sciatic area **with regard to differential diagnosis** from such as depend upon limited, spinal, focal affections, peculiar difficulties will be encountered. Our knowledge concerning the way in which the nuclei of the particular muscles are grouped in the anterior horns of the spinal cord, is, in many respects, still incomplete. At all events, there is a great similarity between these spinal groups and the innervation area of the peripheral nerves. This hint should be sufficient to warn every one to be careful.

In supranuclear paralysis of the leg produced by lesion of the pyramidal tract, the extensors of the foot are usually injured very gravely. This predilection is frequently so remarkable, that the thought of a peripheral complication often suggests itself at once.

Bilateral peroneal paralyses are usually polyneuritic.

Therapy.—We call particular attention to prophylaxis with regard to contractures, which is so very necessary in peroneal paralysis. The difficulty in walking, due to elevation of the heel, may be remedied by means of suitable apparatus, which partially replaces the function of the muscles by elastic straps. For tibialis paralysis, similar apparatus has been invented.

Paralysis of the gluteal nerves is very rare. It has been observed, f. i., after fracture of the os sacrum and in pelvic tumors. The same injury may affect the posterior cutaneous nerve of the thigh. The motor paralysis affects the gluteal muscles and the tensor fasciæ latæ. Extension and abduction in the hip joint are paralyzed; and the inner and outer rotation greatly weakened. When walking, the lack of lateral fixation of the pelvis against

the thigh is especially striking. When stepping on the diseased leg, the pelvis deviates towards the sound side; the movement becomes somewhat waddling.

5. Nerves of the Pudendal and Coccygeal Plexuses

Radicular paralyses of this area will be discussed with the affections of the cauda equina. Next to nothing is known concerning affections in the area of the real plexuses and their peripheral branches.

Supplement

Occupation Paralysis and Occupation Neuritis

As we have seen, *typical paralyses* of single peripheral nerves may be caused by occupational injuries. Thus, continual pressure produces paralyses of the foot, especially of the peroneal nerve, in persons who are forced to work in a squatting position, paralyses of the ulnar in persons who during their work continually support their elbows on something, while pressure and traction produces plexus paralyses in those who carry heavy loads.

One group of cases, however, requires a separate discussion, namely, that in which **peculiar clinical pictures** are occasioned by the special conditions of occupational ætiology, and cases of selective injury of a few known muscles, which in the particular occupation are usually synchronously strained and compressed or otherwise irritated mechanically.

The most important form practically, affects especially workers who must handle continuously a certain tool, the handle of which they must clasp tightly, so that the small muscles of the hand are both strained and exposed to pressure, as ironers, locksmiths, borers, planers, cutters (who must use scissors) and workers in many other occupations. Precisely the same disturbance is seen in persons who are compelled to lean with one hand continuously and heavily on a crutch. In all these cases the muscles of the ball of the thumb and of the interosseus primus are especially endangered.

Occupational paresis (which is similarly localized) of milkers, cigar-makers, etc., may mostly be traced to strain.

We shall not enumerate the rarer analogous occurrences, except the paralysis of a few muscles of the left thumb of drummers (drummer-paralysis), in diagnosing which particular care should be taken not to confound it with a surgical affection which manifests itself under the same circumstances (rupture of the tendon of the extensor longus pollicis, drummer's tendon).

Besides those persons who by reason of toxic or infectious injuries are predisposed to such disease, novices who do their work with an unnecessary expenditure of force, are especially liable to it.

The **clinical picture** which is frequently introduced and accompanied by paræsthesia and pains, is mainly composed of weakness of the affected muscles or atrophy, which sets in either rapidly or stealthily.

R. D. and a slight decrease of sensibility in many cases indicate that the affection is neuritic. Yet myopathic cases possibly occur also.

The **diagnosis** will be easy if proper regard be given to the ætiology. Even disturbances of sensibility and pains, when present, may serve to distinguish several spinal affections having externally some similarity. Occupation paresis may combine with occupation neurosis.

The **prognosis** of the pure cases is usually in all respects favorable.

Of particular importance in the *therapy* is rest for the affected part, and above all, of course, the patient must temporarily or permanently abandon the injurious occupation.

Polyneuritis

Polyneuritis (*Leyden*, about 1880) is a more or less widespread primary degenerative disease of the peripheral nervous system, distributed nearly symmetrically and taking a regular course, and developed fundamentally from a general injury, resulting from toxic matter in the very broadest sense of the word, with which the bulk of the bodily fluids have become contaminated.

Hence not every multiple disease of peripheral nerves is a case of polyneuritis. Such diseases in which a multiple paralysis of the cranial nerves or of the roots is occasioned by gummatous infiltration or sarcomatous tumors in the area of the meninges, of the base of the skull or of the spinal cord, are on the best of grounds not considered as polyneuritis. Nor would multiple paralyses resulting from tumors or gummatous processes on the peripheral nerves themselves come under such classification. Other cases are much more akin to polyneuritis. We have observed before, that upon a substratum of general injuries, which are usually at the bottom of polyneuritis there frequently develop isolated paralyses of peripheral nerves. Paralyses of several single nerves, the irregular choice of which is presumably due to specific local causes also occur rather often under such greatly predisposing conditions. These cases must likewise be distinguished from polyneuritis.

On the other hand, however, local factors, such as heavy functional demands on certain parts, may in some respects modify the distribution even in polyneuritis proper. Nevertheless it will be necessary and almost always possible to separate distinctly, both theoretically and practically, the polyneuritis, the symmetric polyneuritis, from other multiple nerve paralyses (multiple mononeuritis or polyneuritis disseminata and multiple tumor formations).

Ætiology.—The majority of cases of polyneuritis in Germany are occasioned by chronic alcoholism. Next to this the form known as lead paralysis

is probably of greatest practical importance. In the majority of cases chronic poisonings sustained during work with lead occasion lead paralysis, but such diseases have been seen, also, after all kinds of more casual poisonings. Among the classes of workmen who are thus endangered are those who are engaged in the metallurgical extraction of lead; those who cast the metal into shot, pipes, type (type founders), or use it for chemical preparations, such as colors; also painters, varnishers and dyers who use lead colors, potters who apply glaze containing lead, compositors who set lead type; gold leaf workers, filecutters and engravers who use lead plates as supports; workers who must handle the lead plates used in storage batteries, plumbers and others who solder pipes with lead alloys and tinners who use lead for the same purpose.

Arsenical paralysis, after an improper medical use of arsenic, is the next frequent toxic polyneuritis. Polyneuritis from bisulphide of carbon poisoning is seen in some rare cases in workers employed in the vulcanizing rooms of rubber factories. Toxic polyneuritis after poisoning from carbonic oxide, mercury and some other poisons is a great rarity.

Polyneuritis accompanying and following infectious diseases forms a second important group. Of greatest practical interest is undoubtedly the so-called post-diphtheritic paralysis, which most frequently develops about the third week of convalescence.[1] In connection with typhoid, influenza, malaria and other acute infections polyneuritis is much rarer. In these cases the nerve affection also usually sets in during the convalescent stage of the primary disease. In syphilis, polyneuritis manifests itself in the secondary stage, generally not later than half a year after the infection. It also occurs occasionally in the course of gonorrhea and in advanced pulmonary tuberculosis. We shall not enumerate here the rarer infectious causes. In a supplement to this chapter we shall briefly consider the nerve affection in leprosy.

Besides the main ætiological groups of toxic polyneuritis and those appearing in connection with infectious diseases, the so-called cachectic-dyscrasic forms occupy practically only a subordinate place. Those cases occasionally seen in various marasmic conditions and in metabolic diseases, as diabetes, and polyneuritis manifesting itself during pregnancy or when lying-in, unless it can be referred to an accompanying infectious disease (puerperal fever), are classed among the above mentioned forms.

In quite a number of cases polyneuritis has to be designated as idiopathic for want of detectable ætiological factors. It should not be forgotten that even in the toxic, infectious and cachectic-dyscrasic forms the immediate cause of the disease is entirely obscure, for it is only in a small percentage of

[1] The prejudice of the laity that diphtheritic paralysis is a result of the serum treatment has no foundation whatsoever. On the contrary, it seems as if serum treatment instituted very early lessens the danger of an occurrence of this disease. (Heubner.)

those affected with such injuries that a polyneuritis really develops. It is not infrequent, therefore, that even such a predisposing factor can not be found with certainty. Some of such cases give the impression of an independent infectious disease.

A particular, probably infectious, form of disease, endemic in Eastern Asia, Brazil and Africa, which sometimes spreads even into Germany, is called beri-beri.

It remains only to be added, that in the ætiology of polyneuritis several of the factors mentioned may concur and that a cold may in many cases be an exciting factor.

The **clinical picture** of polyneuritis is composed of numerous peripheral nerve paralyses and nerve pareses distributed more or less strictly symmetrically over both sides of the body.

In the following more minute description we begin with a fundamental type, such as for instance in alcoholic polyneuritis and in the majority of the other forms constitutes the rule, and then continue by discussing the clinical peculiarities of some special cases.

In the great majority of cases the nerves of the leg, and among these, the peroneal nerves, are affected first and most severely. Frequently only one leg is affected at first but very soon the other succumbs. Next to the peroneal the tibial nerves are affected, as a rule. Ascending, the disease then seizes upon the nerves of the thigh and the pelvis, and usually together with them, or even before them, also a part of the nerves of the upper extremities, in most cases first the musculo-spiral, then the median and ulnar nerves. In the upper extremities too, a partiality for the distal parts manifests itself. Less often are the shoulder girdle and the musculature of the upper arms affected. The supinator longus muscles of the radial nerve's innervation area are also not infrequently spared. Cases in which the paralysis of the arms bears the character of a plexus paralysis are very uncommon.

As a rule, the paralysis does not develop with equal severity in the various nerve areas. A complete paralysis occurs most frequently in the musculature of the leg. In other nerve areas, it is often only a case of a more or less serious paresis. In grave cases, almost or quite complete, paralysis of all four extremities may indeed occur, and in such cases, the muscular apparatus of the trunk, the abdominal musculature, and the musculature of the back and neck will not often be spared.

Usually it soon becomes possible to recognize the atrophy of the affected muscle-groups. Electric stimulation indicates what changes are to be expected; generally it will be R. D. Almost always the reflexes are very much weakened or destroyed. Destruction of the tendon and periosteal reflexes may be one of the earliest symptoms. Only rarely can a transient initial exaggeration of the reflexes be noted. Even in those light cases in which the tibial and crural nerves are comparatively slightly affected, it is unusual for

the knee jerk and Achilles tendon reflexes to be preserved all through the illness. More often an exaggeration of the cutaneous reflexes (plantar reflexes), particularly in hyperalgesic cases, sets in.

By the paralyses the members are placed in anomalous positions. The feet are in the pes equinus position; the hands usually show wrist-drop, or in some cases, the ape hand or talon position, or a mixed form. In protracted duration of the affection, the anomalous positions are fixed by contractures, unless treatment prevents it.

When the patient is still able to walk, or as soon as he has regained that ability, a pronounced paretic gait, steppage gait or "waddling gait," according to the extent and distribution of the affection, will be observed. Among the earliest symptoms of many cases of disorders occurring in the sensory area, are paræsthesia, formication, and a sense of numbness particularly in the distal parts of the extremities, beginning in the feet. Frequently, but by no means always, very irritating, drawing, burning pains manifest themselves in the parts affected, while the nerve trunks and the musculature are sensitive to pressure. When the pains are severe, and the paralysis is not total, the patient is wont to resist passive movements, which sometimes misleads the inexperienced into thinking that he is dealing with spastic conditions.

Objective disorders of sensation are of far less importance than motor disorders, and even in grave cases sometimes confine themselves to a few nerve areas of the distal parts of the extremity. However, it is very unusual for them to be lacking entirely. Especially in alcoholic and arsenical paralyses a rather extensive hyperalgesia of the skin sometimes occurs, besides the decrease of the other sense qualities commonly restricted within narrower bounds.

Disorders in the secretion of sweat, hyperhidrosis or anhidrosis, reddening of the skin, and a glossy skin, are among the trophic and vasomotor-secretory disorders occurring more frequently. Considerable desquamation and now and then edema are also seen.

The participation of the phrenic and of the cranial nerves is one of the rarer symptoms of polyneuritis. Facial, eye muscle and vagus paralyses in particular, less frequently optic, auditory and other forms of neuritis, are now and then observed.

Very unusual are disorders of innervation affecting the bladder and rectum.

Not infrequently persons affected with polyneuritis show symptoms of a psychosis. A certain symptom-complex of psychic disorders has in fact been designated as polyneuritis psychosis (*Korsakow*). It is undoubtedly true that it is found very frequently in persons affected with polyneuritis. It is particularly characterized by a serious disorder of the ability to receive and retain new impressions. Patients who often can still recollect events of the

more remote past, are entirely incapable of giving a correct account of the events of the immediate past, or even the last few hours. The resulting void in the memory is then filled with "fairy tales" (confabulation). So, e. g., a patient who for weeks had been confined to his bed with paralysis, told the doctor who was making his morning round, that he had already taken a walk in the morning, and had then eaten his dinner with much relish.

Particular Clinical Forms

1. **Pseudotabes Peripherica.**—This form, found especially in alcoholics, but also after diphtheria, in those affected with diabetes and occasionally from other causes, is characterized by the fact that its symptoms of lost function manifest themselves preponderantly in the sensory area. Among the earliest subjective complaints of the patient is a motor ataxia—which has a strong resemblance to the tabetic one—of the lower, later perhaps also of the upper, extremities. This ataxia, like all others, depends in particular on extensive disorders of the deeper sensibility. In such cases, anæsthesias of the skin occur more pronouncedly than in other cases, and are localized corresponding to the ramifications of the cutaneous nerves. The Romberg phenomenon is present; the reflexes are destroyed. Symptoms of motor weakness, on the other hand, play only a secondary rôle, but at least in a few nerve areas (e. g., the peroneal area) they will hardly ever be entirely absent.

2. **Lead paralysis,** in the great majority of cases, and especially as a rule in adults, is confined to the upper extremities. It first affects the musculospiral nerve, usually starting with the right, and going over to the left, in a very peculiar selective fashion, first affecting the extensors of the fingers and those of the wrist. The disease may be restricted to this narrow area. If it progresses, it next seizes upon the long abductors of the thumb and the small muscles of the hand supplied by the ulnar and median nerves. A further extension to brachio-radial, supinator, and other arm muscles, is quite uncommon, while a generalization of the paralysis is a true rarity. A predilection of the paralysis for the lower extremity, in particular, for parts of the peroneal area, is most likely to be found in children. The sensibility almost always remains unimpaired.

This most peculiar picture undoubtedly indicates a spinal rather than a neuritic disease (*Erb, E. Remak*). But the numerous existing anatomical reports do not permit any doubt as to its neuritic character. The peculiar manner of localization is still a matter of controversy.

3. **Postdiphtheritic paralysis** is also distinguished by a peculiar localization. Almost without exception, a motor paralysis, and sometimes an anæsthesia of the soft palate (velum palatinum) is the first and often the only symptom. The speech of the patients is openly nasal; liquids that they are drinking partly flow back through the nose. Frequently a paralysis in

accommodation accompanies the soft palate paralysis, to which it is interesting to note, the hyperopics especially, who read much during convalescence, are said to be predisposed.

In some cases, still other eye muscles, especially the external recti, the muscles of the œsophagus, the vagus branches in the larynx, and other areas of cranial nerves become affected. More frequent is a participation of the extremities. Sometimes its only indication is the destruction of the tendon reflexes of the legs. In other cases, a typical, polyneuritic paralysis of the upper and the lower extremities sets in, and even further generalizations occur. Instead of that, however, the picture of pseudotabes in some cases combines with the palatal and accommodation paralysis.

One is inclined to regard the paralysis of the palate after diphtheria not as a polyneuritic symptom, but as a kind of ascending neuritis, especially as after diphtheria of the umbilicus, a paralysis of the abdominal muscles has been observed. Yet on the other hand, paralysis of the soft palate and paralysis in accommodation have been seen to set in after diphtheritic infections in the arm.

The **diagnosis of polyneuritis** is in most cases easy. Leaving the typical postdiphtheritic and lead paralysis, which, especially with a knowledge of their ætiology, can hardly be mistaken, out of consideration, the following signs of the clinical picture will serve especially to distinguish polyneuritis from other extensive paralyses and paralytic conditions: its symmetrical distribution; its incipiently progressive, as a rule ascending, course which later has a tendency to recovery; the nature of the symptoms of lost function; the paralyses connected with R. D. besides the disorders of sensation which only rarely will be absent; and, lastly, the almost always obvious grouping of disorders corresponding to the innervation area of peripheral nerves. Nor is the psychosis of Korsakow found in those affections which are of any differential diagnostic importance.

We shall now mention the most important special differentiating signs of those diseases, which at one time or another, are most likely to be mistaken for polyneuritis.

In acute poliomyelitic paralysis the progressive course is lacking. It sets in with all its gravity, and then retrogresses. Only in very exceptional cases is its distribution symmetrical. Disorders of sensation are almost always absent. Spinal progressive muscular atrophies are characterized by their more creeping progress, by the lateness at which functional disorders are observed, by fibrillary twitchings which are very unusual in peripheral diseases, and also by the absence of disorders of sensation. Concerning the differentiation (which sometimes becomes difficult) from the so-called neuritic progressive muscular dystrophy, a disease which likewise sets in very slowly and chronically, we refer to the chapter in this work dealing with it.

Genuine tabes is distinguished from the ataxic form of polyneuritis by a number of specific traits, foremost among which is reflex pupillary immobility; but also by other signs, some of which set in very early, such as gastric crises, primary atrophy of the optic nerve, bladder disorders, which are very rare in polyneuritis, and, of course, by the segmental arrangement of the sensory losses. On the other hand, the paresis, which will hardly ever be altogether lacking in typical tabes is absent in pseudotabes. In tabes there often exists an analgesia of the nerve trunks to pressure; in pseudotabes, there is an abnormal sensitiveness to pressure.

Myopathic diseases, which may lead to extensive, likewise symmetrically distributed, disorders of motility (progressive dystrophies, polymyositis, osteomalacial paralysis, myasthenia), lack the symptoms which indicate directly a disease of the nerves, namely R. D., early decrease or destruction of reflexes, and disorders of sensation. We need not mention here how each individually is often revealed at a glance by certain positive signs to those skilled in them. We refer only to the peculiar type of localization in the various forms of dystrophy and to the inflammatory swelling of the muscles, and in polymyositis, frequently of the skin also.

Chiefly, of course, in the interest of successful treatment, a complete diagnosis of polyneuritis naturally requires that the ætiology, founded upon objective criterions, be determined as accurately as possible. Hence it will be necessary to look for objective signs of chronic alcoholism (psychic changes, tremor); of lead poisoning (the blue line on the gums, basophilic granulation of the erythrocytes, lead in the urine, which in some cases will be revealed only after administering iodine); of an abuse of arsenic (cutaneous alterations, melanosis); of CS_2 poisoning (psychic alterations); and for signs of a former or existing infectious disease or metabolic affection.

In the treatment of a diphtheritic paralysis, the frequently occurring myocarditic complication must be taken into consideration. In the disease known as beri-beri, a myocarditis, taking the same course as the symptoms of circulatory weakness, particularly edemas, is really part of the clinical picture.

Course and Termination. Prognosis.—Polyneuritis may set in acutely, even with fever, or may develop slowly. In some rare cases, the disease progresses rapidly from day to day, and soon reaches its highest point of development. More frequently, however, the symptoms require a number of weeks, or even months for their full development. The more chronic development progresses steadily, or, sometimes, by fits and starts. Carelessness of the patient, such as excessive indulgence in alcohol, may at times lead to an exacerbation of the symptoms.

It has already been pointed out in the discussion of symptomatology, that different cases attain to varying degrees of extension. Some, in fact,

remain rudimentary. In the form described as the fundamental type, the affection sometimes does not spread beyond the lower extremities.

The prognosis in general is usually exceedingly favorable.

A fatal issue, caused by the affection alone, is a rare exception. Some cases taking a highly acute and rapidly ascending course, do, indeed, result in death in a short time. Paralyses of bulbar nerve areas, and of respiration, which latter are often of such nature as to be difficult of explanation, frequently cause death. Even in cases taking a less violent course, bilateral phrenic and vagus paralyses may threaten life by their various attendant dangers. It is hardly necessary to mention that for persons whose general condition is wretched, the length of the disease may become serious, or that the gravity of the primary disease may endanger life.

The great majority of cases, as we have already said, recover. Sometimes a complete restoration takes place after a few weeks. Even in widespread cases this occurs, if the paralyses and the alterations of electric irritability do not attain a very high degree. But in most cases, nevertheless, it takes many months or even longer in grave cases, to recover. Lasting defects may usually be avoided.

Naturally much depends on the possibility of completely eliminating the causal injuries. Alcoholics who do not renounce alcohol, those affected with lead poisoning who continue to receive the poison into their system, cannot hope to recover.

Only a few words concerning the **pathological anatomy** are necessary to supplement the discussion in the general part. It is a matter of a primary degenerative process, a parenchymatous neuritis. The peripheral ramifications of the nerves are more likely to be seriously affected than the large trunks and roots. It seems that the fibre disease, which is typical of polyneuritis, is the degenerative form beginning in the periaxial medullary sheaths. Slight changes of a like nature are frequently found in nerve areas, which have not given evidence of any symptoms of lost function (latent neuritis). The unequal, intermittent extension of this destructive process makes it possible that Wallerian degeneration sets in in a fibre segment which at first remained normal, when the change in the neighboring proximal parts has progressed to the destruction of the axon. One is inclined to explain in this manner the discovery of fibres which show the Wallerian degeneration type, in seriously diseased nerves. In cases of long standing numerous fibres disappear wholly (Figs. 38 and 39).

The changes in the connective tissue of the nerve, as a rule, appear to be secondary. Pure inflammatory focal affections of the nerve branches and trunks are found now and then, but they are always quite circumscribed in extent and are an inconstant accompanying symptom, showing nothing typical.

In the spinal cord and spinal ganglia are often found those slight changes,

closely related to peripheral nerve diseases, which have been discussed in the general part. In passing we may remark that besides these, complicating focal affections of the brain and spinal cord may sometimes occur in some forms of polyneuritis, as inflammatory and other cerebral focal affections in diphtheria, and the so-called encephalopathia saturnina in lead poisoning.

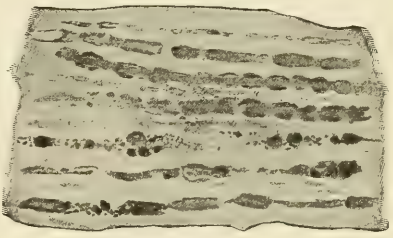


Fig. 38.—Fresh polyneuritic degeneration. (*After Bälz and Miura.*) Teased preparation of the peroneal nerve in a case of beri-beri. Osmium staining. In some fibres pronounced periaxial medullary disintegration; in others apparently Wallerian degeneration.

The **therapy** of polyneuritis requires but a brief discussion, since the important general rules which apply to polyneuritis as well, have been dealt with at length in the discussion of the paralyses of the single nerves. Because of this we may pass over the requirements of absolute rest in new cases, of a suitable diet, of proper position, in connection with which especially the prophylaxis of passive contractures should be considered and of other requirements.

In order to treat the causal conditions properly, we should insist on total abstinence in alcoholics, eliminate the poison in those affected with lead poisoning by administering iodide of potassium (2 to 3 times daily 0.5 (5 grs.) in solution), and treat malaria with quinine, diabetes with a proper diet and syphilis energetically with mercury. Any previous inunctions should not be regarded as interfering with this, unless, indeed, symptoms of mercurial poisoning should exist. Some cases which in spite of the treatment continue to progress in the beginning, will often recover completely in a remarkably short time if the treatment is continued longer.

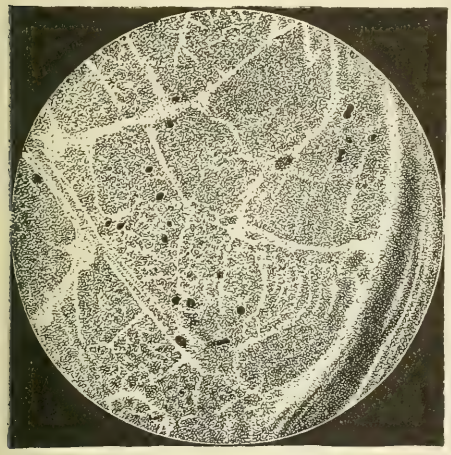


Fig. 39.—Polyneuritis alcoholica in a later stage. (*After Jacob.*) Cross-section of the n. ischiadicus. In the proliferated connective tissue only a few isolated (black colored) fibers are preserved.

No definite rule can at present be established concerning the difficult question of interrupting pregnancy in pregnancy polyneuritis (cf. von Hösslin, Arch. f. Psychiatrie, vol. xl, and Münchner med. Wochenschr., 1905, page 636).

For the rest, the use of moist, warm packings of the whole body, or of the diseased parts is to be recommended in the early stage. They may be applied daily from 1 to 2 hours and will almost invariably prove beneficial,

particularly in painful cases. Suitable for every stage—in the earliest for those patients, at least, who are not too sensitive—are thermal baths of 96° to 98° F., continued on the average for half an hour, taken 3 to 6 times a week. Sometimes medicinal preparations (pine needle extract, salt, carbonic acid, etc.), may be added. Frequent diaphoretic procedures such as hot air baths in the bed, are also urgently recommended for both the early and later stages of the disease (*Oppenheim*).

The tonic pills mentioned before should be used rather than other medicinal remedies, to aid the general treatment.

For the pains, antineuralgic agents or, if necessary, also the stabile anodic galvanization are used besides warm local applications.

When the affection does not progress any further, and especially during convalescence, the other electro-therapeutic means, massage, and passive and active gymnastics should be used according to the rules mentioned above. Careful passive movements may sometimes prove advantageous even in the earlier stage.

As after cures, thermal bathing resorts furnished with good medico-mechanical institutions are particularly to be recommended.

Careful treatment will obviate the necessity of having to remove any residual defects by means of orthopedic surgery.

Naturally any grave special occurrences during the course of the disease will require special treatment. In deglutition paralyses it is better to have recourse to artificial feeding than to risk an aspiration pneumonia. In grave acute disorders of respiration, the life of the patient may be saved by artificial respiration (Ebstein).

Supplement

A. Landry's Paralysis, Paralysis Ascendens Acuta

As Landry's paralysis (Landry, 1859) all those cases are commonly designated, in which a severe and flaccid paralysis begins acutely in the feet, thence progresses rapidly and steadily to all remaining muscles of the legs, trunk, arms, neck and the bulbar nerve areas. In sporadic cases the onset of the disease is marked by a febrile attack. The reflexes are destroyed and usually there are slight disorders of sensibility. As a rule, no real pains are experienced. Disorders of bladder or rectum are found only exceptionally. Death generally results in the first few days or the second week (rarely later) from suffocation or respiratory failure. Some cases recover slowly.

A descending course of the disease has also been described.

It is undoubtedly true that a polyneuritis acutissima may occasion this clinical picture. But it would be wrong, simply to identify Landry's paralysis with polyneuritis. Some postmortem findings seem to indicate that it may also be caused by an ascending myelitis. Besides there are some cases in

which a remarkable condition of electric irritability exists which is incompatible with either polyneuritis or myelitis. Autopsies very rarely afford a satisfactory explanation of the clinical picture.

At any rate it is a matter of a toxic or infectious disease. It may appear in connection with certain infectious diseases and toxic processes. Swelling of the spleen and albuminuria may follow in its wake. In not a few cases micro-organisms of various kinds have been found in the tissues of the body, particularly in the central organs.

It is at present impossible to distinguish clearly from each other and to classify the cases of Landry's paralysis.

B. Leprosy of the Nervous System

Leprosy of the nervous system is now and then very erroneously simply designated as polyneuritis leprosa.

Grave disorders of cutaneous sensibility, in its earlier stages, as a rule, extending irregularly in spots, or appearing in segmental radicular zones and afterwards covering extensive areas, even the whole body, sometimes affecting all qualities, sometimes only one or more, stand in the foreground of the clinical picture (lepra anæsthetica). The sensibility of the deeper parts usually remains unimpaired, for which reason ataxia hardly ever occurs. The anæsthesias combine with muscle atrophies, usually only in limited areas, among which there is a predilection for the small muscles of the hand, and with grave "trophic" disorders, which partly depend undoubtedly upon the nerve disease and partly constitute symptoms of local leprous changes leading even to loss of entire finger joints (lepra mutilans). The state of the reflexes is always irregular. Typical paralyses of peripheral nerves appear only in exceptional cases.

It is obvious that this clinical picture may be mistaken for syringomyelia more easily than for polyneuritis, despite the fact that both halves of the body may be affected almost symmetrically. It may be very difficult to distinguish it from syringomyelia. But usually the characteristic spotted and nodular leprous changes of the skin will be found in leprosy of the nervous system, while in syringomyelia a few symptoms, like the spastic palsy, will manifest themselves, which as a rule are not found in leprosy.

The pathological-anatomic explanation of the picture of "lepra nervorum" is not at all conclusive as yet. It is certain that alterations of minute nerve branches of the skin together with leprous cutaneous diseases play a rôle in it. But besides these, there are also, perhaps ascending, bacillary invasions of the larger trunks, the spinal ganglia and the spinal cord, with or without consecutive, especially interstitial, changes (neuritis interstitialis). Occasionally lepra nodules on the larger nerve trunks destroy them. Just how often primary polyneuritic degeneration of the nerves

occurs in such cases as a result of a general toxæmic disposition, can not be definitely ascertained.

II. THE NEURALGIAS

Neuralgia is used to denote a pain, appearing paroxysmally and spreading along the course of a peripheral nerve. That it is a matter of an anatomically limited disease of the peripheral nervous system, cannot be doubted, since it appears in such a localized manner.

The pathologic-anatomic fundamental elements are by no means definitely known. It may be assumed that it is a matter of irritating influences affecting the nerve in some point or other of its course. In some forms, indeed, as we shall see below, pathologic changes of the nerve or in its vicinity are known, which may cause the irritation. How and when the characteristic attacks take place after the nerve is affected by an apparently permanent injury, is still undetermined.

The nerve trunks themselves, as seen under the microscope, are in many cases entirely normal. The slight diffuse neuritic changes which occur in some cases, are probably only in exceptional instances related directly to neuralgia. As a rule, neuritis does not manifest any genuine neuralgic symptoms. On the other hand, slight neuritic symptoms may often appear, even clinically, in neuralgia.

Ætiology

In many cases neuralgia follows *infectious diseases*. Probably the most frequent one in Germany is that which appears in the area of the supraorbital nerve during or after influenza. In persons who have suffered from malaria, attacks of neuralgia may take place at the same typical intervals at which the attacks of fever appeared (masked malaria). Secondary and tertiary syphilis may lead to neuralgia; in the tertiary form it is usually produced by perineuritic or paraneuritic gummatous processes.

Now and then neuralgia, especially the form of it accompanying herpes zoster, appears under a clinical picture of an independent infectious disease, and even, according to some reports, as small endemics.

Toxic processes do not play an important part in neuralgia. Very prominent, however, is the predisposing significance of *metabolic diseases*, foremost among which is diabetes mellitus.

Colds are frequently given as causes of neuralgia. It is especially difficult then not to recognize such connection between the two, when that part of the body which was affected by the cold becomes the seat of neuralgia (sciatica after sitting on the cold floor, etc.).

Of special interest are those cases in which a *chronic irritation of a certain part of the nerve trunk* may be directly proven. We mention tumors in the

broadest sense of the word, osteitis and periostitis, bone fragments, scars and inflammatory processes in the neighborhood of the nerve. Tumors of the nerve itself may be disclosed by true neuralgia, especially in the early stage. Diseases in the area of the nerve roots, the vertebræ and meninges must be considered.

The pathogenic irritation may proceed also from the *peripheral extension area* of the nerve. Various inflammatory processes, cicatrices and many painful alterations of a chronic nature may occur. In amputation neuromata the patient often imagines that he feels the pain they occasion in the part that has been removed.

According to an old theory neuralgia may be excited *reflexly* from a distance. Thus it is said that neuralgia may develop in a distant nerve area from the intestines because of intestinal worms; from female genital organs because of a reflexio uteri, etc. This, however, is very questionable.

Furthermore *transient mechanical irritations* of a nerve may result in a neuralgic condition, though the pathologic connection can not be explained in detail. To this class belong neuralgia after contusion of the affected part of the body and postpartum sciatica.

Finally, *severe functional strain* upon some parts plays a certain rôle. So, a supraorbital neuralgia is often seen after any work in which the eyes were strained and a long continued one sided strain upon one leg will result in a sciatica of that leg.

Lastly we mention that *general debility* seems to develop a tendency to neuralgia.

Neuralgias are distinguished as *symptomatic and idiopathic*. All those cases in which a local or general fundamental disease may be found are called symptomatic. As long as it is impossible to find such disease, the neuralgia is called idiopathic. It will be noted that this classification is not based on any important internal difference, and is really worthless. Strictly speaking, neuralgia is always only a symptom.

The practical result of this, as we shall point out also later, is the necessity of looking carefully for a palpable cause in every case.

Such a conception also makes it unnecessary to separate from neuralgia pains of a true neuralgic character manifesting themselves in other organic nerve diseases, such as basal cerebral tumors, meningitic processes.

Clinical Picture

As a rule, the attack of pain is preceded by a brief premonitory paræsthesia. It very soon attains its highest point. Pains have been described as splitting, burning, boring, cutting and pricking, and very frequently they are intolerably violent. Asked for the seat of the pain, the patient will often indicate with his hand or finger the exact course of the diseased nerve. In grave or protracted neuralgia, an irradiation frequently takes place, so that

the whole vicinity of the nerve begins to pain also. Neuralgia of one nerve may be accompanied by pains in its secondary branches and its neighboring nerves.

The attack may pass away in a few minutes or last for hours.

The attack sets in either spontaneously or is excited by certain external causes, among which are various sensory irritations, particularly those that affect the diseased area, as touch, shock, draughts of air, thus movements and exertions of the diseased area, for instance eating in trigeminal neuralgia, walking in sciatica, and finally coughing, pressure, mental excitement and changes in the weather.

Especially in chronic cases, and most frequently in sciatica, slight pains are often either constant or nearly constant, and the attacks appear then as temporary more or less pronounced exacerbations.

Of foremost diagnostic importance among other signs of the disease are the so-called pressure points (*Valleix*, 1852). These limited points of exceeding susceptibility to pressure are found in places where the nerve has been slightly irritated mechanically, where it rests exposed upon a hard foundation, where it emerges from bony canals or where it pierces a fascia. The tenderness to pressure persists usually in the intervals between the attacks; rarely during the latter alone. It is said that it may even be lacking entirely. Slight pressure on the pressure point is liable to excite an attack, while strong pressure is often soothing. Less often the entire course of the nerve is tender to pressure. *Trousseau* regarded as very important the regular susceptibility to pressure of certain spinous processes in different neuralgias.

The disease is further accompanied by vasomotor, secretory and trophic disorders. Reddening and a slight swelling of the skin in the area of the pain is frequently seen during the attack and finally may become habitual. More extensive cutaneous changes are often occasioned by mechanical mistreatment, to which many patients expose the affected parts during the attack. Occasionally hypersecretion of glands situated in the area of the injured nerve, especially a surplus flow of tears and saliva in neuralgia of the fifth nerve are seen. Among trophic disorders anomalies of hair-growth occur. By far the most important is herpes zoster, which will be discussed briefly in a supplement to this chapter.

In the motor area, tonic tension and clonic spasms of the muscles in the affected parts are sometimes seen during the attack of neuralgia. A tic convulsif may combine with the "tic douloureux," in trifacial neuralgia. The great care which patients bestow upon the affected member, occasions a kind of pseudo-paresis. The muscles of the affected area become more or less wasted in all graver and more protracted cases. As a rule, however, true pàreses or paralyses do not occur, except in a minority of cases in which a local progressive process forms the basis of neuralgia, which in time leads to destruction of the motor nerve fibres.

Sensory disorders are frequently represented by hyperæsthesias of the skin. Slight hyperæsthesias in peripheral neuritic and sometimes in radicular areas, also occur often.

Weakening or loss of a reflex, such as the Achilles tendon reflex in sciatica, is in grave cases very common.

It is hardly necessary to mention that the general condition of the patient may be greatly injured by the disease.

But little remains to be added concerning the **special conditions of a few quite important single neuralgias.**

Trifacial (*trigeminal*) *neuralgia*, aside from sciatica, is by far the most frequently occurring form. Usually only one branch, the first in most cases, is affected. The most important pressure points are situated at the supra-orbital foramen for the first branch, at the infraorbital and zygomatico-facial foramina for the second, and at the mental foramen for the third. It is probably only in syphilitics that a bilateral neuralgia of the auriculo-temporal occurs, the pain area of which, in the form of a child's comb, extends across the crown from one ear to the other, and whose pressure point is situated on the zygomatic arch in front of the ear (Seeligmueller). In neuralgia of the first branch, the pain usually extends along the ramus supraorbitalis. In the area of the second branch a predilection for the rami infraorbitalis and alveolaris, next to the zygomatic nerve is shown, in the area of the third branch, for the ramus alveolaris inf.

Among the causes, the affections in the extension area of the nerve, such as diseases of the nose and its cavities and especially of the teeth, play a special part particularly in trifacial neuralgia. Even the sharp edge of the shrunken alveolar processes in persons who have lost their teeth may excite a neuralgia.

Hence the advice of a dentist rarely can be dispensed with. A random extraction of sound teeth, such as has been observed occasionally, is of course wholly inadvisable.

If in herpes zoster trigemini the cornea is affected, the aid of an oculist is to be sought.

In *occipital* neuralgia, which chiefly affects the major occipital nerve, and which belongs to the frequently bilaterally occurring neuralgias, tumors of the posterior cranial fossa and affections of the uppermost vertebræ are ætiologically to be considered. The main pressure point is situated on the linea nuchæ sup. at the point where the nerve comes to the surface through a hiatus in the muscle.

True *brachial* neuralgias are very rare. They are most likely to occur (either uni- or bilaterally) in affections of the meninges or vertebræ and in tumors in the root or plexus area. In an ætiological search cervical ribs must also be sought for.

Intercostal Neuralgia.—Among special ætiological factors are affections of

the ribs, various intra-thoracic diseases, space contracting processes such as aneurisms and severe scoliosis, although, of course, the pains in the chest occasioned by all these diseases are only now and then of a truly neuralgic character. The pressure points are situated in the intercostal space near the vertebræ, in the mid-axillary line and near the sternum.

A neuralgic affection of the nerves of the mammary glands which appears especially after trauma during pregnancy and lactation, is designated as *mastodynia* (irritable breast, Astley Cooper).

Neuralgias of the lumbar plexus nerves, neuralgia lumbo-abdominalis, cruralis, etc., are very rare.

Sciatic Neuralgia (Ischiadica).—Sciatica, the malum Cotunnii, (Cotugno, 1764) is the most frequent form of neuralgia. Among the special causes are diseases of the os sacrum and pelvic bones, tumors and exudations of the pelvis, displacements of the uterus, sciatic hernia and chronic constipation. A manual examination of the pelvic organs, through the rectum, should never be omitted in any case of sciatica.

In bilateral sciatica there is always a suspicion of a meningeal or vertebral affection. Next to that, it is probably most likely to occur in diabetes.

Sciatic neuralgia may be restricted to single branches, like the posterior cutaneous.

The pressure points are situated at the great sacro-sciatic foramen, in the gluteal crease, in the popliteal space, in the middle of the calf, at the head of the fibula, behind the ankle, sometimes also laterally from the tubercles of the os sacrum, somewhat below the middle of the crest of the ilium and in other places.

Important for the diagnosis is the pain when stretching the nerve. Flexion of the hip joint when the knee is in extension excites violent pains, while no pains are occasioned if the same movement is executed with the knee flexed (phenomenon of Lasègue).

The patient is apt to carefully avoid all movements which might produce such stretching and he likewise avoids exposing the back part of the leg, and through it the nerve, to any pressure. Most patients keep the leg slightly flexed at hip and knee both when lying and walking and rotate it somewhat outward. When standing or walking they throw the weight of the body on the healthy leg. The trunk is slightly lowered towards the healthy side (sciatic scoliosis). In some rare cases, however, a slight deviation of the spinal column in the opposite direction takes place. The origin of the scoliosis is still very doubtful (Figs. 40 and 41).

The reflexes of the affected leg are not infrequently slightly increased. It has already been mentioned that sometimes they are also weakened. Occasionally a peculiar kind of "pseudo-Babinski" reflex is found, whose nature has not yet been explained.

The **diagnosis** of a typical neuralgia is easy, if strict regard be given

to the main symptoms, which have been mentioned in the definition, the anatomic extension and the paroxysmal character and course of the pain. Difficulties ought to arise only in atypical forms.

When the course as to the paroxysms is not clearly pronounced, as occurs in cases of long standing and especially also in sciatica, the other symptoms, particularly the localization of pain and perhaps also the anam-

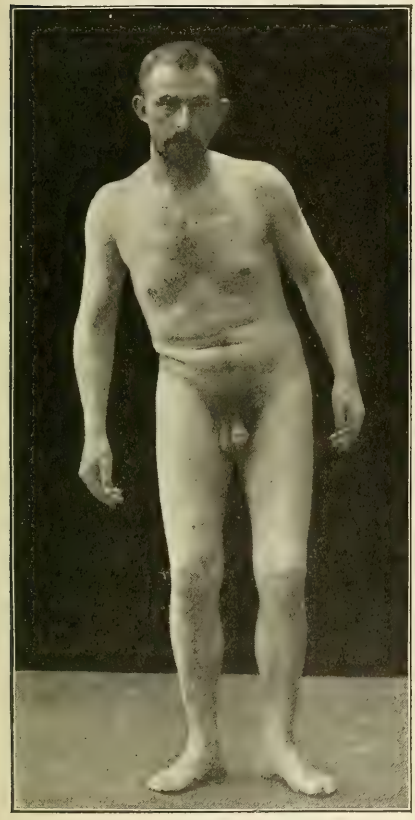


FIG. 40.

FIG. 41.

FIGS. 40 and 41.—Sciatica at the left side. Characteristic posture of the leg, scoliosis ischiadica. (*Leipsic Medical Clinic.*)

nestic statement that characteristic attacks occurred before, will frequently permit of a correct diagnosis.

Where the localization in the course of a certain peripheral nerve cannot be authenticated, especially in neuralgoid conditions, in which the pain is confined to a very narrow area, it will as a rule, be impossible to come to any definite conclusion. This is true for instance of the so-called joint neuralgia, achillodynia, mastodynia, coccygodinia and visceral neuralgias.

In all doubtful cases it should be ascertained whether other, especially inflammatory affections, do not cause the existing pain. As we have seen,

the existence of such a disease, of an inflammation, or of a tumor would not necessarily exclude the existence of neuralgia. The latter may exist independently of such affections, but, on the other hand, may not only be unfavorably influenced by them, but even caused by them. It will therefore always be a question of whether criteria of neuralgic pains are present or whether the disorders correspond simply in nature and course to the other disease which has been found.

We shall now mention a number of diseases, which as experience has taught us, are frequently, with but a superficial examination, mistaken for neuralgia. Thus neuralgic pains are easily confounded with the acute and radiating pains in diseases of large joints of the extremities, especially of the hip and shoulder joints, in hereditary deformities such as flat-foot, in affections of the arteries and veins (intermittent claudication, varicose veins), and in occupation neuroses. The so-called lymphangitis rheumatica (Wilms[1]) must also be considered.

Catarrh of the frontal sinus and even common attacks of migraine have occasioned a wrong diagnosis by being mistaken for trifacial neuralgias and beginning pleurisy has been confounded with intercostal neuralgia.

Of a more general, differential diagnostic significance are muscular rheumatism, tabetic pains, spasms, neuritis, those reflex pains and hyperæsthesias in visceral affections which have been investigated mainly by Head, to which perhaps belong the pains in angina pectoris, and psychogenic pains.

In all these painful diseases the characteristic signs of neuralgia, aside from those rarer cases which are combined with a true neuralgia, are of course lacking. We can not here deal with their positive symptoms.

The differential diagnostic significance of psychogenic pains is considerably enhanced by the fact that in some cases, particularly in individuals with a nervous predisposition and in those who because of injuries claim insurance money, they may add themselves to true neuralgia and strongly resemble it. At any rate, it may be seen in such cases how the type of the neuralgic picture becomes more and more obliterated, while the signs of the "psychalgia" (Oppenheim), the determination of the form and course of the disorders by certain ideas, the dependence upon attention and expectation, becomes more and more pronounced and exclusive. Perhaps in those cases of neuralgia, which ostensibly have been cured by hypnotism, it was a matter of such psychogenic residual images.

Once the presence of neuralgia has been conclusively determined, the investigation should continue as to the ætiology. The general condition of the body should be carefully considered; one should search for infectious and toxic conditions and for metabolic disturbances. Next, those parts through which the diseased nerve takes its course, should be closely scruti-

[1] Münchener med. Wochenschr., 1906, page 1595.

nized, to see whether any alterations exist which might sustain or produce the irritated condition of the nerve. Above all it should be discovered whether some grave, progressive, local disease is not hidden behind the neuralgia, such as a malignant neoplasm or something of the sort. Thus, for example, a tubercular meningitis may begin under the innoxious picture of a sciatica. Bilaterality, which is not infrequent, rise in temperature, the proof of other tubercular diseases or of a tubercular diathesis, may early confirm one's suspicion.

The **prognosis** should always be made with caution, even in those cases where it is believed that a graver fundamental disease does not exist. The prospects are comparatively favorable in many post-infectious neuralgias. A neuralgia following influenza may recover in a few days. Diabetic neuralgia often disappears after one succeeds in removing or at least considerably diminishing the glycosuria. In the majority of cases a neuralgic affection lasts a number of weeks or a few months. Even its continuation for several years is by no means uncommon.

Old age, a poor general condition of the patient, a long course and particularly the severity of the disease all darken the prospects for recovery. In many cases the tendency to relapse is quite marked.

Therapy.—On account of the many sided ætiological relations of neuralgia great care is required in its treatment. The general condition should be improved as far as possible. Any existing morbid disorders, particularly those, of course, that from experience are known to have been factors in the origin of neuralgia, should be taken up for treatment. After that one should endeavor to remove the local causes.

Abstinence from alcohol is almost invariably to be advised; the use or non-use of coffee or tea must be decided upon individual grounds in each case. Experienced authors warn against a one sided diet, unless it is very definitely indicated. Regulating the bowels is of great importance in all cases; in a few rare cases it has actually direct curative value.

Particularly in recent cases it is advisable to keep the affected parts in a condition of absolute rest for some time. Recent cases of sciatica necessitate confinement to bed for two or three weeks, longer if the condition of the patient requires it. It will sometimes be advisable, to secure rest for the diseased parts in a way most comfortable to the patient by means of splints or by other fixation methods.

From the abundance of various remedies we have selected the following important ones, which have stood the test of time.

Among the *medicinal agents*, quinine, iron, mercury and the iodides play their well known rôle in the specific ætiological treatment of malaria, anemia, syphilis and lead poisoning.

A general treatment with tonics, particularly arsenic and strychnine, and with the ubiquitously tried iodides is believed to be useful in many cases.

The antineuralgics occupy a foremost position. In recent cases, a regular use of several (say about 3) doses daily is advised, afterwards they may be kept in reserve to be used when necessary. Of the great number of these agents we mention antipyrine (0.5 to 1.0 per dose); phenacetine (0.5); antifebrin (0.25); migrainine or antipyreticum compos, Riedel (1.0); pyramidon (0.3); lactophenin (0.5); aspirin (0.5 to 1); quinine sulfat. or muriat. (0.5). When the single preparations do not prove successful, mixtures may often prove of value, such as antipyrine 0.5, phenacetine antifebrin āā 0.25, 1/2 powder for one dose. Combinations with bromine and with codeine are to be recommended. As another agent adapted to alleviate pain, we mention butylchloral. We caution against the use of morphine, in view of its uncertain action, and the danger of morphinism, since the duration of the disease can not be foretold. In extreme cases single doses of morphine or its derivatives (heroin, etc.), will have to be given, just as one will sometimes have to resort to a soporific (chloral hydrate). In some exceptional cases, atropine is found to be soothing where morphine has failed.

Hydro- and Thermo-therapy.—Of the general applications in recent, not very sensitive, cases warm baths of half an hour's duration, or hot baths for a shorter time, taken several times a week should be considered, also the more energetic, diaphoretic hot air baths in bed (they may be taken even by the most seriously affected patients), by means of such apparatus as that of Hilzinger, of 120° to 140° F., and about 1/4 to 1/2 hour length. Later, especially as after-treatment, thermal baths, carbonic acid and brine baths may be recommended.

Local thermic procedures are usually applied upon the painful places and preference will almost always have to be given to warm or hot applications. Moist or dry, warm or hot bandages, cataplasms, thermophores or similar applications, packing in peat or mud, sand baths, hot air baths, most conveniently in Bier's apparatus, radiations from photo-therapeutic apparatus are successfully prescribed, those most convenient to the patient in the beginning, the others in the more advanced stages of the affection.

From the group of the so-called derivative agents which have stood the test of experience, we recommend especially dry cupping glasses, to be used even in recent cases and applied daily along the course of the affected nerve in number corresponding to the extension of the painful area. Embrocations which irritate the skin, tincture of iodine applied with a brush, plasters which produce cutaneous irritations and which may be left on for several days (emplastr. cantharid. perpet. and empl. oxycroceum, etc.), applications of menthol spirits and menthol oil under covering compresses, perhaps sometimes even real vesicants (emplastr. cantharid. ordin.) may all be used to alleviate the pain. Blood letting, applications of moxæ, etc., should be resorted to but seldom.

The chief method of electro-therapy is the stabile anodic galvanization of the main points of the pain.

A button shaped electrode (surface 5 to 10 qcm.) is placed on the pressure point; a larger one, the cathode, is placed as the dispersing electrode on any point in the healthy area, as for instance, in sciatica in the region where the nerve originates (os sacrum region). In order to attain success it will be necessary, when beginning and ending the treatment, to increase and diminish the current very carefully and to avoid all irritating influences. Weak currents of about 2 ma., when the nerve is situated deeper, 3 to 4 ma., should be used. Seances to last from 2 to 5 minutes. Occasionally more protracted applications may be tried. At the same sitting various pressure points, each for about 2 minutes, may be treated successively. Galvanic treatment is given from 3 to 7 times a week; in exceptional cases even twice daily. If applied correctly the result will not infrequently prove a direct palliation of the troubles.

Besides this preeminently successful method, a great number of others have been recommended. Particularly in inveterate cases, for instance of sciatica, it may be expedient to place both electrodes (about 10 qcm. in size each) proximally and distally on the affected nerve and to let somewhat stronger currents (about 5 to 6 ma.) flow through it for about 5 minutes. During this treatment the position of the electrodes may be changed. It seems to make no difference whether the "ascending" (cathode proximal) or the "descending" current is used.

Faradism is not as valuable as galvanization. In the lighter cases of long standing faradization of the skin and muscles of the affected area is sometimes beneficial. The treatment of painful places by means of the faradic brush should be tried only in exceptional cases.

Mechano-therapy.—In inveterate cases, particularly of sciatica, massage of the affected parts is exceedingly serviceable. Rubbing and kneading may be followed by vibratory massage of the affected nerve along its entire exposed course. In recent cases massage almost invariably proves a failure.

Stretching the nerve, which today is probably used only in the form of so-called bloodless stretching, may be appropriately combined with massage. For example, in sciatica after the leg has been massaged and is in extension at the knee it should be lifted from its support very slowly, so as to occasion as little pain as possible and then be flexed in the hip joint more and more. Avoiding all rough manipulations one should proceed as far as the patient can bear it, then keep the leg in that position for 1 to 2 minutes, and then perhaps after a final, stronger stretching, let the leg down slowly. Manual stretchings of aching cutaneous areas are recommended also, and have undoubtedly been used in some cases, in which they afforded an instantaneous palliation of pain (Naegeli).

Climatic therapy is of no considerable importance. In general, very changeable, irritating, cold or moist climates should be avoided.

Of the many methods of injection of different liquids into the nerve trunk or its immediate vicinity we recommend chiefly a trial of the method given by Lange, which is almost entirely painless. It consists in a paraneuritic injection, repeated when necessary, of rather large quantities (about 100 ccm. in sciatica) of a solution of eucaine-β in a physiological solution of cooking salt (cf. Münchener med. Wochenschrift, 1904, No. 52, page 2325). This causes a rapid palliation and even stopping of the pain, and in some cases leads to a rapid and permanent recovery.

Among the surgical methods, which are discussed more fully elsewhere in this volume, neurolysis, releasing the nerve from its irritating surroundings and neurinsarcoclesis may be ranked among the treatments removing the cause. The mutilating operations of neurotomy and neurectomy, of neurexairesis and of extirpation of ganglia, can be advised only in those cases in which the resources of internal medicine have been exhausted. An operation should not be performed before careful galvanic treatment, which even in cases of old standing often rewards us with surprising results, has been applied by an expert. Even surgical methods do not insure a permanent cure.

SUPPLEMENT

Herpes Zoster

Herpes zoster consists of an eruption accompanied by pain. Quite rapidly there develop one after another groups of vesicles on a reddening surface. These localized groups have a highly characteristic lengthwise formation in a stripe-like area, corresponding to the innervation area of a certain nerve root, or sometimes of a peripheral branch. The affection which is almost invariably unilateral, most frequently occupies the area of an intercostal nerve, a somewhat beltlike region on one side of the thorax, whence its name. Next to this, the trigeminal area is most often affected. Sometimes the vesicles disappear after a few days, leaving either brown spots, which later disappear, or, if the efflorescences were hemorrhagic or gangrenous, white scars typically grouped.

Anatomically, with the greatest regularity a disease of the peripheral sensory neuron, especially of the spinal ganglia, but not infrequently also of the peripheral nerves, and frequently true inflammatory changes, are found. The real relation between the cutaneous affections and the nerve alterations is entirely obscure.

Frequently, though by no means invariably, signs of a greater suspension of function and anæsthesia occur during herpes zoster. Motor paralyses, like

facial and eye-muscle paralyses in herpes zoster trigemini, are symptoms that rarely occur.

Many traits of which we mention only the frequent initial fever, the occurrence of epidemics, and the immunity which from the rareness of relapses may be supposed to exist, all indicate that in numerous cases a peculiar infectious process forms the foundation for this clinical picture. Among the causes of herpes zoster, there are a number of factors which we have also found in the ætiology of neuralgia, namely: other infectious diseases, affections of the vertebræ, trauma, etc.

FIG. 42.

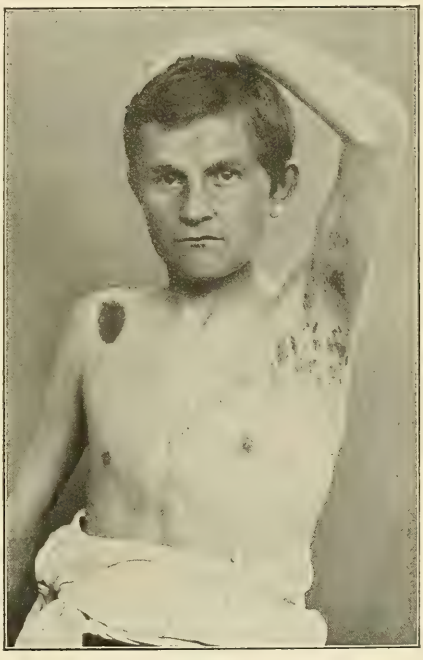


FIG. 43.

FIGS. 42 and 43.—Herpes zoster. Fig. 42, in the region of one of the middle thoracic nerves. Fig. 43, in the region of the second thoracic nerve. On the shoulder at the right a birthmark, (*Leipsic Medical Clinic.*)

Herpes zoster is closely related to carbon oxide and arsenical poisoning. Occasionally herpes zoster manifests itself in connection with diseases of internal organs, by appearing in a neighboring cutaneous zone, for instance in inflammations of the costal pleura. The connection between the two has not yet been explained. In many cases of herpes zoster, no definite ætiological factors can be found.

The relation of herpes zoster to neuralgia can probably be summed up thus: that they stand in a sort of co-ordinated position to each other. The anatomic changes of the nerve which form the basis of the cutaneous affection, in many, but not all cases, give rise to neuralgia. It may be that the

disorder occasionally is not accompanied by a cutaneous affection, and that it becomes apparent only as a result of a neuralgia.

Zoster neuralgias form but a comparatively small percentage of all the cases of neuralgia.

Neuralgia may precede herpes zoster for some time, may begin in association with it, or may follow it.

No difficulty is, as a rule, experienced in distinguishing zoster from other forms of herpes. In treating the eruption one may generally confine oneself to a light protective bandage, some dusting powder, etc.

III. THE TUMORS OF THE PERIPHERAL NERVOUS SYSTEM

As we have seen, tumors seizing a nerve metastatically or by spreading thence from a neighboring organ, may lead to its neuralgic affection or paralysis.

Neuromata, the tumors emanating primarily from the peripheral nerve, may occasion the same clinical picture. The special symptoms of these tumors require a separate discussion.

It is relatively rare that neuromata emanate from the nervous tissue proper. It is usually a matter of so-called spurious neuromata, tumors that emanate from the connective tissue of the nerve and which histologically appear as fibromata, lipomata, or even as angiomata. Some tumors may degenerate and become malignant.

We shall consider first those forms appearing *singly or* at any rate *confined to a certain, small area of the body*. They develop either on the larger trunks or on the fine terminal ramifications, particularly in the subcutis. The latter group includes the so-called plexiform (Ranken) neuromata, which are almost exclusively of merely cosmetic surgical interest, and the small, innoxious nodules, designated as tubercula dolorosa. These are often very dense, sometimes angiomatously livid and painful. They appear singly or in small groups and may be easily removed by an operation.

The neuromata of the trunk not infrequently lead to neuralgia and paralysis. Intra-cranial neuromata of the cranial nerves, as they appear, especially on the auditory and trigeminal, give the same clinical picture as the basal brain tumors.

Neuromata on the intra-vertebral spinal cord roots produce the symptoms of extra-medullary spinal tumors.

Besides this form of solitary neuromata there exists one of multiple neuromatosis. The most frequent form of the latter is the general one of the skin and subcutis. Fibromata or lipomata, softer or harder nodules, are generally found, sometimes in enormous numbers, distributed over the whole surface of the body. The neurofibromatosis of the skin is designated also as Recklinghausen's disease (after Recklinghausen 1882). In some cases,

however, the tumors of the large nerve trunks, of the cranial nerves and spinal roots appear in large numbers. Occasionally numerous neuromata are found in the skin besides the single or several centrally situated tumors of this nature, and the determining of these may become very valuable for a correct diagnosis of any central tumors appearing under the disease picture of a spinal or brain tumor. At any rate, the most varied forms and localization of neuromata may be found together in one and the same individual: besides nodules on the nerve trunk, appearing in a wreath like formation, root and skin neuromata, multiple tumors in the deeper part of the body, in the muscles and the intestines, so that one may indeed speak of a general neuromatosis.

The disease is evidently based upon a congenital predisposition. It may appear even very early or, on the other hand, only late in life. It occurs in whole families. The persons affected with it often show other signs of a morbid disposition and are affected with deformities, numerous nævi, pigment anomalies, etc., and show a neuropathic diathesis.

The clinical importance of the disease varies with the number, size and location of the tumors. Especially the neurofibromatosis of the skin may be almost devoid of troublesome features. In other cases they may become very annoying through acute pains. Some neuromata situated comparatively indifferently, may become dangerous through excessive growth. The danger of sarcomatous degeneration in multiple neuromatosis and in solitary neuroma of the trunk is rather great.

Aside then from the entirely harmless, or nearly so, local affection of the surface of the body, the prognosis is very uncertain. There are cases of a mild nature, entirely lacking in any propensity to propagate, but there are others also with a strong tendency to growth on the part of the existing tumors, and a disposition to form more and more new ones. Strange to say, a spontaneous retrogression of single tumors has also been reported.

Surgical treatment should be used if seen to be absolutely necessary. When advising patients one should always remember, however, that in such cases as well as in all others, mechanical irritation may cause the transformation of harmless tumors into malignant ones.

III

DISEASES OF THE SPINAL CORD

1. NORMAL AND PATHOLOGICAL PHYSIOLOGY OF THE SPINAL CORD

BY

MAX ROTHMANN (Berlin)

The **spinal cord** represents phylogenetically the oldest apparatus of the central nervous system. In the lowest vertebrate, the amphioxus, it represents therefore, since the brain is quite rudimentary, a virtually independent central apparatus, which shows normal functions, even after the removal of the head-end. In the ascending scale of vertebrates, the dependence of the spinal cord on the various higher centers of the brain increases continuously, and special investigation is needed in the highest mammals, to still find individual independent functions of the spinal cord. It is therefore readily understood, that the spinal cord in building up its own centers, and its connections with the brain, furnishes us means to recognize anatomical and physiological arrangements peculiar to itself in each and every species of animal. Comparative anatomy as well as physiology, therefore, may transfer only with the greatest caution, the facts gleaned from one species of animal to another however nearly related. This should be especially considered in making use of the result of animal investigation for the purposes of human pathology.

The spinal cord lies in the vertebral canal. At the upper end, it is rather arbitrarily bounded by the exit of the first cervical nerve and the lower boundary of the crossing of the most important cerebro-spinal tract, the pyramidal crossing. At the lower end, in adult man, it is far from reaching the bottom of the vertebral canal, but as a rule reaches only the lower part of the first lumbar vertebra. In lower mammals and in the early fœtal life of man, on the other hand, it occupies the entire cavity of the lumbar and sacral canal. While the spinal cord in its development thus remains behind the growth of the vertebral column, it is replaced in the lumbar and sacral vertebræ by a delicate thread, the *filum terminale.*

By this absence of the spinal cord in the lower part of the vertebral canal of man, lumbar puncture invented by *Quincke,* at the level of the lower lumbar vertebræ, has been made possible without danger of lesion to the spinal cord.

The spinal cord is enclosed by the *spinal membranes.* On the outside lies the *dura mater,* a comparatively wide sac, which at the lower end fuses

with the filum terminale. On the inside of the dura lies the *arachnoid* which, as a fine network, fills the space between the dura and the pia, and contains the cerebro-spinal fluid, and the *pia mater*, lying directly upon the surface of the spinal cord and sending branches into its fissures. The spinal cord is usually divided into *cervical*, *dorsal*, *lumbar and sacral* portions with the end in the *conus terminalis;* this latter, by gradual attenuation, goes over into the filum terminale. To this corresponds the division of the *spinal cord roots* into 8 cervical roots, the uppermost of which emerges from the vertebral canal between the occipital bone and the atlas, whereas the lowest emerges between the 7 cervical and the first dorsal vertebræ, into 12 dorsal, 5 lumbar and 5 sacral roots, the lowest of which leaves the vertebral canal between the os sacrum and the os coccygis, and the coccygeal nerve.

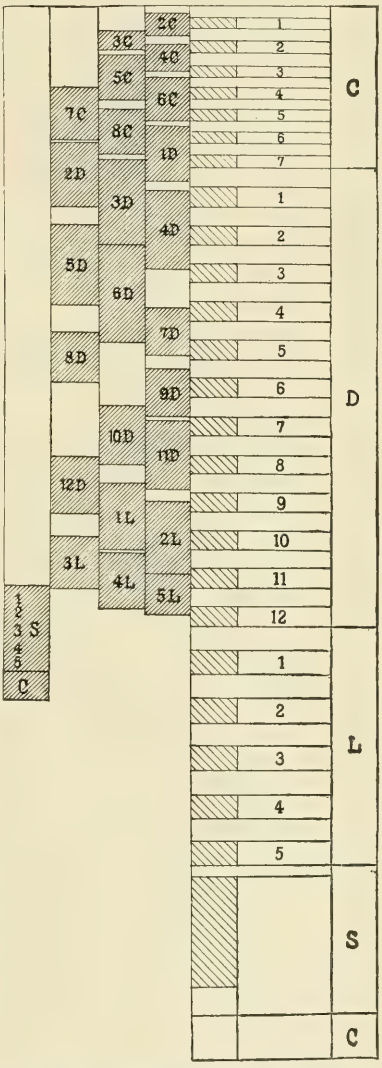


FIG. 44.—Scheme of location of the individual spinal-cord segments in the vertebral canal. (*After Reid.*)

The above mentioned gradual shortening of the spinal cord in relation to the vertebral canal, leads to a *displacement of the spinal cord segments* upwards, so that the individual spinal cord roots, in order to reach the foramina of exit destined for them between the vertebræ, have to course some distance downwards. The length of the lumbar and especially of the sacral roots, is considerable; the downward coursing lumbar and sacral roots form, around and below the conus terminalis, the cauda equina. The relation between the spinal origin of the individual roots and their places of exit from the intervertebral foramina, which is of the greatest importance for spinal cord localization, appears clearly in Reid's diagram (Fig. 44). In the dorsal portion of the cord, the entry of the root into the cord lies about 2 to 3 vertebræ above the corresponding intervertebral foramen of the vertebral canal, but in the lumbar and sacral roots this difference increases with extraordinary rapidity; so that, for instance, the sacral roots all have their spinal origin at the height of the twelfth dorsal and first lumbar vertebræ.

In the spinal cord, there are *two rather pronounced enlargements*, corresponding to the origin of the roots of the extremities, the *cervical enlargement* from about the third cervical to the first dorsal vertebra and the *lumbar enlargement* from about the tenth to the twelfth dorsal vertebræ. Both appear in fœtal life synchronously with the development of the extremities.

In the spinal cord the *gray substance* is everywhere surrounded by *white substance*. Only the tips of the posterior horns touch the periphery at the place of entry of the *posterior roots*. The changing formation of the white and the gray substance decides the different forms seen in transverse section of the spinal cord in the various segments. Immediately below the pyramidal crossing, the gray substance, by development of the anterior and posterior horns, assumes the characteristic butterfly figure. The *anterior horns*, the seat of the great motor cells, are, naturally, developed especially well in the cervical and lumbar enlargements. But the development of the gray substance in the entire lumbo-sacral portion of the cord, absolutely as well as in relation to the quickly diminishing white substance is a particularly strong one. In the dorsal portion of the cord a process, which may also be observed between the anterior and posterior horn at other points of the cord, the reticular process is developed into a *lateral horn*. The posterior horn shows considerable thickening where it passes into the medulla oblongata, as well as in the lower segments of the sacral portion of the cord. At its base lies *Clarke's column*, developed especially well in the dorsal portion of the cord; the head of the posterior horn follows, then the peculiarly transparent gelatinous substance of Rolando, the marginal zone and the apex of the posterior horn, which is to be regarded as belonging to the glial covering of the cord. Both symmetrical halves of the gray substance are bound together by an anterior and a posterior *gray commissure*, which embrace, both in the front and the rear, the *central canal* with its surrounding central gelatinous substance. In front of the anterior gray commissure lies a broad bridge of white substance, the *anterior white commissure;* but numberless medullated fibres pass also through the gray commissure.

The *white substance of the spinal cord* is divided into posterior, lateral, and anterior columns. Whereas the separation between the posterior and lateral columns is rather clearly indicated by the apex of the posterior horn and the entering bundles of the posterior roots, no definite anatomic boundary can be established between the lateral and anterior columns; the ventral border of the roots which is, however, frequently interrupted may be considered as the boundary. Between the two anterior columns lies the deep *anterior fissure*, which reaches the anterior white commissure that connects the anterior columns. Between the two posterior columns is the *septum posterius*, usually not quite opened into a fissure; it reaches the posterior commissure. Between the septum posterius and the dorsal line of roots, which deepens into the sulcus lateralis posterior, there is mostly a sulcus

intermedius posterior, which permits us to divide the posterior column into a lateral part, the *column of Burdach*, and a medial part, the *column of Goll*.

The *spinal cord roots* are divided into anterior and posterior. The *anterior roots* which spring from the anterior horn cells of the spinal cord, pass, separated into several fibres, through the antero-lateral column to the periphery, where they unite into a compact bundle. The *posterior roots* spring from the *spinal ganglia*, which are situated in the intervertebral foramina, outside of the dural sac. Only the lowest spinal ganglia, and, exceptionally, those of the uppermost cervical roots, lie intradurally in the vertebral canal. Hence the posterior roots pass to the sulcus lateralis posterior, in order to penetrate the so-called entry zones of the roots of the posterior column. While the anterior roots are thus placed ventrally to the spinal ganglia, the anterior and posterior roots unite laterally from the ganglion into a bundle, the peripheral nerve.

Concerning the *distribution of the blood* in the spinal cord, every *vertebral artery*, before union with the basilar artery, sends a branch backwards, and, higher up, one to the front. Both anterior arteries, A. vertebrospinales anteriores, unite, running downwards, in the top part of the spinal cord, into an azygos artery, which, in the cervical enlargement, terminates in the tractus arteriosus spinalis anterior. The latter is formed of an anastomotic chain of the anterior short branches of the intercostal, lumbral and sacral arteries, down to the filum terminale. From this, the central arteries pass through the anterior fissure to the anterior commissure and send each a branch into the gray substance of either side; only the peripheral divisions of the latter are also supplied with blood from other sources. Furthermore, from the tractus arteriosus spinalis anterior, lateral branches reach the anterior roots, the anterior and antero-lateral column region.

The posterior vertebro-spinal artery unites with the posterior anastomotic chain of the intercostal, lumbar and sacral arteries to form the tractus arteriosus posterolateralis, one on each side. From its medially passing rami penetrantes, a second tractus arteriosus posterior is formed medially from the posterior roots. From all these vessels, some arteries pass into the white substance, the vasocorona, and supply this as well as the peripheral parts of the gray substance. All arteries entering the spinal cord are terminal arteries.

The veins of the spinal cord, upon whose course we can not enter here with greater minuteness, are more developed than the arteries; they are all characterized by the absence of valves. Though we know very little of the course of the lymphatic vessels, their product, the *cerebro-spinal fluid*, has, in recent years, attracted attention in a constantly increasing degree. This clear fluid of about 1003 specific gravity fills the subarachnoid space as well as the cavities of the central nervous system; it shows a slightly alkaline reaction, contains small quantities of globulin, and about

1 % of solid substance. *Quincke's* lumbar puncture, the cocainizing of the spinal cord, introduced by *Bier*, and in most recent times, the serodiagnostic syphilis reaction of *Wassermann-Neisser-Bruck* have more and more clearly shown the importance of this fluid, which continually moistens the central nervous system.

So far as the *microscopically* recognizable structure of the spinal cord is concerned, it consists, like that of the entire central nervous system, of *ganglion cells, nerve fibres and neuroglia.* Among the ganglion cells the large *cells of the anterior horn* (anterior root cells) appear as the type of the motor ganglion cell. They are multipolar and have an axis cylinder process, and possess a centrally situated nucleus with nucleoli. The axis cylinder process becomes continuous with a medullated nerve fibre, which passes ventrad into the anterior column. In the cell protoplasm are found *Nissl's granules* (tigroid bodies), among which an achromatic substance is found, the main components of which are the *neurofibrils;* these traverse the body of the cell, and appear in all processes, even in the axis cylinder. The neurofibrils probably represent that part of the body of the cell, which conducts the stimulation, though nothing has been definitely ascertained as to the relations of the ganglion cells to the extracellular fibril structures. One thing, however, seems to be proved by the most recent investigations into the coursing of the fibril, namely, that the theory of the neuron—that is, the entity of the ganglion cell with its axis cylinder, which is said to have no connection with other neurons except by contact, a theory which has been formulated under the influence of *Golgi's* methods of metallic impregnation of the cell—is not tenable from the *purely anatomic* point of view. On the other hand, we must consider the neuron theory for the present as especially valuable for physiological and clinical investigation, and adhere to it because of its great importance for further researches.

In the ganglion cells, especially of the anterior horns, there is found also a peculiar *yellow pigment*, which in adults, as a rule, is found collected in clusters on one side of the protoplasm. This pigment, described as lipochrome because of its behavior to osmium and the fatty extractive substances, is entirely lacking in the newly born. It appears in the child at the age of from 6 to 8, and increases in quantity with advancing years, until in old people of from 80 to 90 it often occupies almost the entire protoplasm of the cell. Since this phenomenon appears with increasing years also in animals (dogs, horses, monkeys) it might be regarded as a product of age, which inhibits the activity of the ganglion cells.

The anterior horn cells are found at the various levels of the spinal cord, in varying numbers, sizes and arrangements, but as a general rule, four groups of ganglion cells may be distinguished by cross-section, a ventromedial and a dorsomedial, a ventrolateral and a dorsolateral. The latter two are very strongly developed, especially in the cervical and lumbo-sacral

portions of the cord. Of other groups of cells, those in the lateral horn and in the intermediate zone between the anterior and the posterior horns are to be mentioned—the *sympathetic nuclei.* According to the most recent investigations, there are three such nuclear columns, the nucleus sympathicus lateralis superior from the eighth cervical to the third lumbar segment, the nucleus sympathicus lateralis inferior from the second sacral segment to the coccygeal portion of the canal and the nucleus sympathicus medialis inferior, from the fourth lumbar segment to the coccygeal portion of the canal.

Besides these, *Clarke's columns* stand out preeminently. This group of cells, situated at the base of the posterior horn, dorsolateral from the central canal, is most strongly developed in the dorsal and lower cervical portions of the cord, but is well marked also in all the other portions of the cord. Clarke's columns are characterized by rather large ganglion cells with large nuclei and nucleoli, relatively few and coarse-grained tigroid bodies, and strikingly numerous protoplasmic processes. We cannot here enter into the structure of the rest of the ganglion cells, which in great variety are distributed in the spinal cord.

The gray matter, as it appears especially in Weigert's hæmotoxylin preparations, is filled by great masses of coarse and fine medullary sheath nerve fibres. Among them we find a relatively small number of fibres running from above downwards; they run for but a short distance. With the exception of the accessorius roots in the upper portion of the cervical segment, which run through the lateral column, only the *anterior root fibres* leave the spinal cord from the gray matter. Besides these, there arise in the gray matter of the spinal cord fibres which pass up and down; they course in the lateral and anterior columns, and also, but fewer in number, in the posterior column. Into the gray matter pass fibres from the various columns of the white matter and fibres of the posterior roots; they radiate, in part directly, and partly as collaterals of the posterior column fibres, into the posterior horn and Clarke's column, but, a portion of them also may be traced to the region of the anterior horn cells. Besides these there are numberless commissural fibres, which, passing through the gray commissures and especially also through the anterior white commissure, connect the two halves of the gray matter; they also make possible a connection of the white spinal cord tracts with the gray matter of the other half of the spinal cord.

Finally relative to the *neuroglia*, which shares with the ganglion cells an ectodermal origin, one must distinguish the ependymal cells, lining the cavities of the central nervous system and therefore also the central canal, and the neuroglia cells distributed throughout the gray and white matter, which, however, seem to have the same embryonic origin. Intermingled with the neuroglia cells, which we must be careful not to confuse with ganglion cells, especially in pathological relations, there exists an extraordinary

fine meshwork of glia fibres. These glia fibres, however, are not to be regarded as independent formations, but as processes of the glia cells. In every loss of the actual nerve substance in the spinal cord, there is a proliferation of the glious elements which leads to the formation of sclerosis.

If we now consider the white matter of the spinal cord, it consists, besides the supporting substances, exclusively of nerve fibres, which serve to carry intraspinal and cerebro-spinal impulses. The separate bundles of nerve fibres bound together for the conduction of special functions, are therefore called *conducting paths*. Since observation of the normal sections of the spinal cord could give but little definite knowledge as to the course of such tracts, the *secondary degenerations*, which appear in consequence of an interruption of certain tracts in focal affections of the central nervous system, were early used to investigate this matter. After a medullated nerve fibre is completely divided, there appears in the peripheral part, separated from the parent cell, swelling, degeneration and finally absorption of the medullary sheath. The fresh degeneration of medullary sheaths may be proved with great accuracy by means of *Marchi's* method, in which osmic acid stains newly degenerating medulla black; the absence of medullary sheaths in older processes can be readily demonstrated by Weigert's medullary sheath method. Only in later stages of the processes, and under peculiar circumstances, is there, besides this cellulifugal degeneration of the peripheral nerve stump, also a retrograde degeneration of the central nerve stump in addition to characteristic changes in the nerve cells themselves.

Another successful means of acquiring knowledge as to the individual conducting tracts of the spinal cord, is *Flechsig's method for the study of the development of the medullary sheath*, which is based upon the disparity in time of the medullary development of the individual tracts. Then comes *Gudden's method of developmental inhibition*, which rests upon the fact that after destruction of a center or a conducting path in a newly born animal, i. e., in incomplete development, all the centers and tracts that belong to the same system fail of complete development.

Finally the *experimental and physiological* methods of irritation and destruction of certain centers or conducting tracts in the grown animal are important for the establishment of the conducting tracts.

Of the *conducting tracts of the spinal cord*, the longest known is the *pyramidal tract* (cortico-spinal tract). It has its origin in the region of the central convolution of the cortex of the cerebrum, and, according to the most recent investigations, predominantly in the anterior central convolution. But the region of its origin must in any case extend over the area of the Betz giant pyramid cells. After its known course through the internal capsule, the basis of the cerebral peduncle, and the pons, it reaches as the pyramid, the ventral surface of the medulla oblongata, and, where this makes its transition into the spinal cord, enters the pyramidal crossing, in such a way

that the fibres from every pyramid pass backwards and cross one another at an acute angle, in the form of bundles, like the fingers of folded hands, ventral from the central canal. From the crossing the pyramidal fibres pass

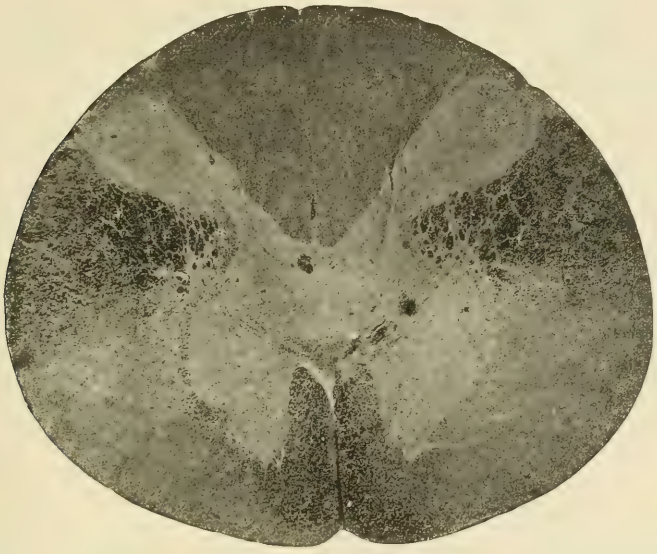


FIG. 45, *a*.—Degeneration of both pyramidal tracts of the lateral columns in the first cervical segment, directly below the destroyed pyramidal crossing in the macacus rhesus. Descending degeneration in the medial parts of the anterior columns. Marchi preparation.

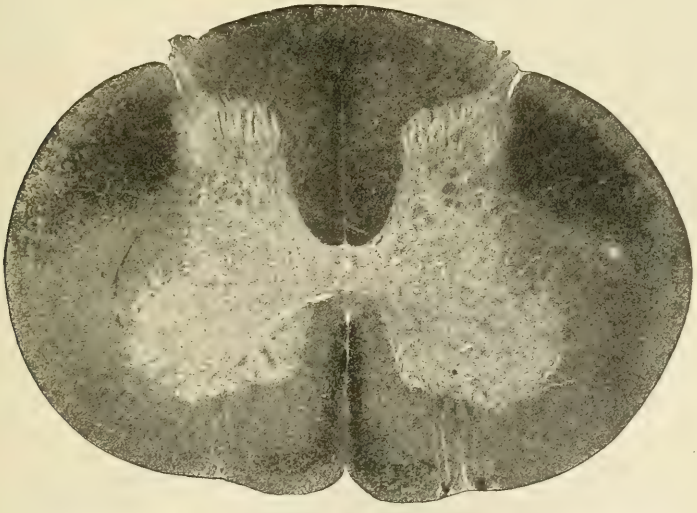


FIG. 45, *b*.—The same degeneration in the seventh cervical segment.

to the base of the posterior horn, and then, with a sharp bend outwards, into the lateral column. While this crossing in the dog and even in the lower monkeys is complete, so that the pyramidal tract connects each hemisphere of the cerebrum directly with the opposite lateral column, there is, in the

anthropoid ape and especially in man, a considerable part of the pyramidal tract, that takes no part in the crossing, but passes uncrossed downwards along the anterior fissure into the anterior column. The relation of the crossed part of the pyramidal tract (*lateral pyramidal tract*) to the uncrossed (*anterior pyramidal tract*) is very changeable in man, so that cases in which the anterior tract is almost lacking are found, as well as isolated cases, in which the pyramidal crossing is utterly lacking and the entire pyramidal tract reaches the anterior column uncrossed. Besides these two kinds of fibres, there are a small number of pyramidal fibres, which, uncrossed, reach the lateral column on the same side; these appear to be somewhat more numerous in man than in monkeys and dogs, in which they may be occasionally altogether absent (Fig. 45, *a* and *b*).

The great variability of the pyramidal tract in man coincides with the ever changing situation and size of this tract in animals, and points to the

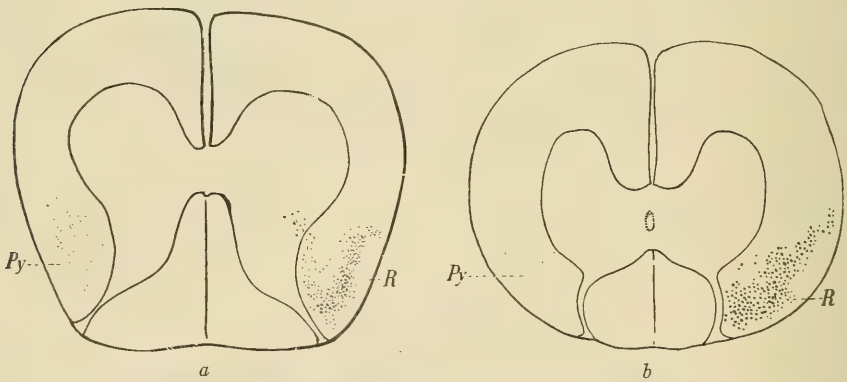


FIG. 46.—Pyramidal tract and rubro-spinal bundle in the spinal cord of a dog. *a*, Cervical enlargement; *b*, dorsal region. *Py*, pyramidal tract. *R*, rubro-spinal bundle.

comparative youthful age of this tract in the animal kingdom. Birds do not have a pyramidal tract at all; it is only indicated in the upper part of the cervical portion of the spinal column in the hedgehog, the sheep and goat; in mice, rats and squirrels it does not pass into the lateral column, but into the top of the posterior column, and we can recognize in passing from the rabbit, by way of the cat and dog, to the monkey, a gradual growth and increase of calibre in the fibres of the tract, which in that species is situated exclusively in the lateral column. Only in anthropoids does a clearly defined anterior pyramidal tract exist.

The development of the pyramidal tract is also relatively late; its axis cylinders begin to appear in man in the sixth to seventh fœtal month. The medullary envelopment does not take place until after birth. The lateral pyramidal tract lies in the postero-lateral column; in the cervical portion of the cord it is separated from the periphery by other masses of fibres; in the lumbo-sacral portions, it is adjacent to the periphery. From it fine col-

laterals pass to the base of the anterior horns, though no one has, as yet succeeded definitely in establishing the exact place where these fibres terminate in the gray matter. At any rate, they do not seem to connect directly with the cells of the anterior horn. The lateral pyramidal tract may be traced downwards into the lowest part of the sacral portion of the cord by staining the tissues by Marchi's degeneration method; it gradually decreases in volume as it proceeds downwards. The anterior pyramidal tract, as a rule, does not extend beyond the lowest part of the dorsal cord.

In about the same area as the lateral pyramidal tract, there runs a second tract descending from the brain to the spinal cord, the *rubro-spinal tract (Monakow's bundle)*. Its main mass springs from the red nucleus of the corpora quadrigemina region, crosses immediately after its origin in Forel's tegmental crossing, and reaches, after receiving some pontile fibres, the postero-lateral column, in which it passes down to the sacral portion of the cord; it sends out comparatively large collaterals to the gray matter at the base of the anterior horns (Figs. 46 and 47).

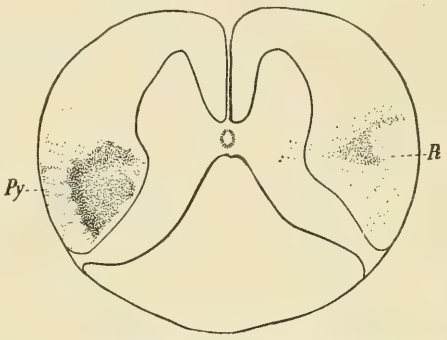


FIG. 47.—Pyramidal tract and rubro-spinal bundle in the ape. Third cervical segment.

This tract stands in inverse relation to the pyramidal tract; in the absence of the latter it is well developed, and as the other increases in the animal kingdom it gradually diminishes. In the dog, the pyramidal tract is still surpassed by it in size, surrounding it laterally and ventrally; in the monkey it may be correctly designated as a prepyramidal column, since the bulk of its fibres is being forced out ventrad by the strong development of the pyramidal tract. In man, it is only a rudimentary prepyramidally situated little bundle of fibres. In about the same area there are also some fibres, which descend from Deiter's nucleus to the spinal cord.

In addition to the anterior pyramidal tract, which is situated alongside the anterior fissure, the anterior column also contains other important tracts passing from the brain to the spinal cord. The largest of these has its origin in *Deiter's nucleus*, a large group of cells lying in front of the cerebellum at the level of the acusticus-nuclei, and reaches, uncrossed in the main, the anterior and antero-lateral columns of the spinal cord, whence, adjacent to the periphery, it passes to the lowest part of the sacral portion. This *vestibulo-spinal or Deiter's tract* is connected by large collaterals with the anterior horn of the same side.

A second important connection between the midbrain and the anterior column is the *corpora quadrigemina anterior tract;* it arises from the gray matter of the anterior quadrigeminal body, crosses completely in *Meynert's*

tegmental crossing, reaches the anterior column of the spinal cord by way of the posterior longitudinal bundle, where, by a wealth of collaterals, it radiates into the anterior horn. This tract is paralleled by a *pontile anterior tract*, rising in the gray matter of the pons; it likewise passes downwards in the peripheral divisions of the medio-anterior column (Fig. 48).

In the most lateral portion of the anterior column, and above all in the ventrad antero-lateral column of the upper cervical portion of the cord, the bundles of fibres course, which descend from the *respiratory centers* in the medulla oblongata to the phrenic centers in the fourth cervical segment, but their course is not exactly known.

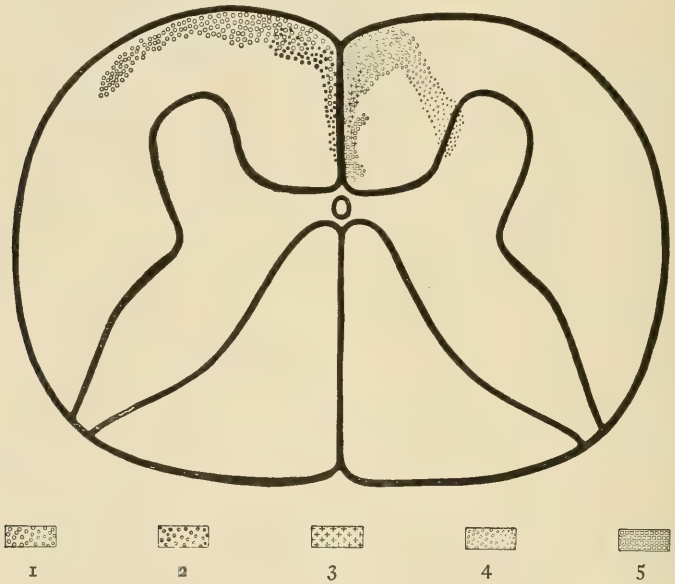


FIG. 48.—Descending cerebro-spinal anterior column tracts in the dog. 1, Tract from Deiter's nucleus; 2, fibres from the pons to the anterior column on the same side; 3, fibres from the pons to the anterior column of the opposite side; 4, tract from the quadrigeminal bodies to the anterior column; 5, fibres of the posterior longitudinal bundle.

In addition to all the tracts here described, which pass from the brain into the spinal cord and degenerate descendingly after they are interrupted, and which occupy, as a whole, peripheral divisions of the spinal cord, there are in all parts of the anterior and lateral columns other intra-spinal *connection fibres*, which pass through the white matter, for shorter or longer distances, in order to connect higher with lower spinal segments. The shorter of these pass on the very margin of the gray matter, while the longer ones approach the periphery and partly commingle with the cerebro-spinal fibre systems.

Now as to the *ascending systems of fibres, passing from the spinal cord to the brain;* there is no such system, which analogous to the pyramidal tract, ascends directly to the cerebral cortex. From the posterior roots, springing

from the spinal ganglion cells, only one tract passes in the spinal cord, without interruption, up to the medulla oblongata: the tract of the *long posterior column fibres*. But only a small part of the posterior root fibres that pass upwards reaches the posterior column nuclei of the medulla oblongata, whereas a large part of them partly in the segment of the root entry, partly in higher spinal segments, unites with the cells of the posterior horns, and Clarke's columns. As the bundle of fibres, entering one segment of the posterior column, places itself along the medial margin of the posterior horn and is then pushed mediad by the posterior root fibres of the next higher segment, there arises an imbricated structure of the long posterior root fibres in the posterior column, in such a way that finally in the upper cervical portion of the cord the posterior root fibres, coming from the posterior roots of the lumbo-sacral portion and almost the entire dorsal portion, are placed in the medial part of the posterior column, the so-called *Goll's column*, and end in the medio-posterior column nucleus of the medulla oblongata, the nucleus funiculi gracilis, while the laterally situated *Burdach's column* is supplied by the posterior root fibres of the cervical and upper dorsal portions of the cord, and ends in the nucleus funiculi cuneati of the medulla oblongata (Fig. 49). None of the fibres of the posterior column goes higher than this; only by the help of the lemniscus tract, rising in the posterior column nuclei of the medulla oblongata and passing to the optic thalamus, and by the thalamo-cortex connections is an indirect connection with the cortex of the cerebrum established.

This posterior column portion of the posterior roots passing directly to the posterior column nuclei of the medulla oblongata, increases gradually n the ascending scale of animals, and is most developed in man.

In addition to the large ascending posterior column tract, every posterior root sends off, at its entry into the posterior column a small *descending branch*. It consists of a strand of fibres, which may be traced downwards a few segments from the root entry; at the same time it gradually moves away from the posterior horn, and attenuates very quickly.

Besides these *exogenous or root fibres* of the posterior column, there exist also *endogenous* fibres, originating in the gray matter of the spinal cord. Our knowledge of these we owe partly to experimental destruction of the gray matter in the lumbar portion of the cord, as it may be produced by anæmization in consequence of temporary occlusion of the aorta abdominalis or by embolization of the terminal arteries of the gray matter; partly also to the study of fresh cases of poliomyelitis, in which only the gray substance is affected in the more or less extended process of inflammation. In such lesions of the gray substance of the lumbo-sacral portion of the cord, one sees at the level of the lesion the ventral top of the posterior column, the *cornu-commissural zone*, filled by moderate degeneration. Hence the field of degeneration extends as a dorsal endogenous posterior column field, in

an upward direction, dorsad along the posterior fissure, and reaches, with a small part of its fibres mixed with the posterior root fibres of the sacral portion of the cord, Goll's nucleus of the medulla oblongata (Fig. 50).

Descending degenerating posterior column fibres, described as endogenous systems, have become known, more especially through pathological observations. These form *Schultze's comma shaped tract* at the border of Goll's and Burdach's columns in the cervical and upper part of the dorsal portion, *Flechsig's oval field* at the posterior fissure in the lumbar portion and the *medio-triangular tract* in the dorsomedial angle of the lower sacral portion of the cord. These fields in their course from above downwards seem to merge into one another, but whether they are purely endogenous descending posterior column fibres, or are mixed with posterior root fibres, has not been definitely ascertained.

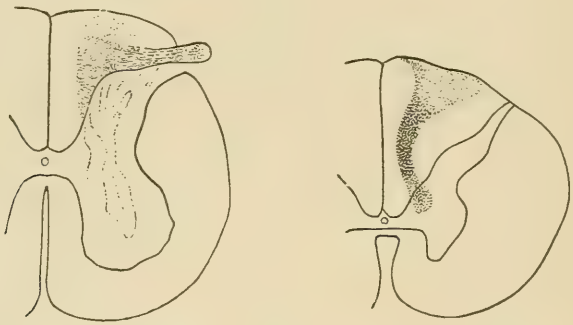


FIG. 49, *a* and *b*.—Degeneration of the posterior column after total destruction of the fifth lumbar root. *a*, Upper part of the fifth lumbar segment; *b*, twelfth dorsal segment. (*After Schaffer.*)

Direct posterior root fibres end only in the posterior columns. *The connection of the posterior roots with the ascending systems of lateral and anterior columns results exclusively* through the mediation of the ganglion cells of the gray matter. Most important here, are the *spino-cerebellar tracts of the lateral column*, which are divided into a *dorsal or Flechsig's tract*, and a *ventral tract or Gower's bundle*. The *dorsolateral cerebellar tract* springs from the ganglion cells of Clarke's column, in the dog from the sacral portion on throughout the entire spinal cord, in man only beginning with the lumbar enlargement; it lies laterally from the pyramidal tract and the rubro-spinal bundle, along the periphery of the dorso-lateral column and extends to the apex of the posterior horn. In its course upward, it sends off numerous little fibres into the gray matter, at points but reaches the medulla oblongata as a compact bundle, enters the inferior cerebellar peduncle, and terminates, mostly crossed, in the superior vermis of the cerebellum.

While this tract was first demonstrated by *Flechsig* on the basis of medul-

lary sheath development, the *ventrolateral cerebellar tract, Gower's bundle,* owes its discovery to the degeneration method. Its origin in the spinal cord is not quite definitely known; but the ventral cells of the posterior horn of both sides seem to send fibres to it. It lies in the spinal cord in the ventral half of the lateral column along the periphery, dorsad directly touching the dorsolateral cerebellar tract, but mediad pressing farther into the white matter of the lateral column in the form of a triangle. This tract, also, constantly receives fibres in its course through the spinal cord, and sends other fibres off to the gray matter. The main bulk of these fibres diverge in the medulla oblongata from Flechsig's tract where this tract enters the inferior cerebellar peduncle; it describes a peculiar curve over the trigeminal root at its exit and over the superior peduncle of the cerebellum, thus reaching the dorsal division of the vermis in which it ends, partly crossed and partly uncrossed.

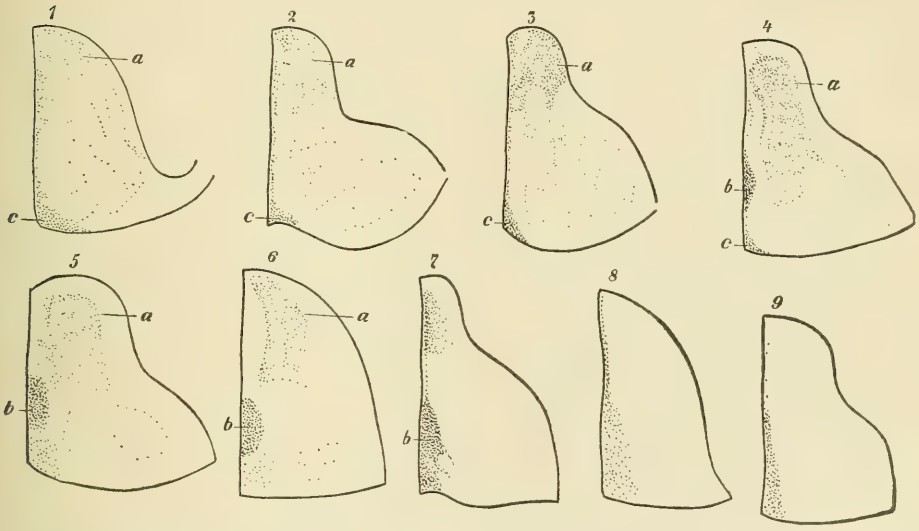


FIG. 50.—General view of the ascending endogenous degeneration of the right posterior column after cutting off the blood supply to the gray matter of the sacral and lumbar cord through lycopodium embolism in the dog. 1, Sacral cord; 2–5, lumbar cord; 6–8, dorsal cord; 9, cervical cord. *a*, Endogenous ventral region of the posterior column; *b*, endogenous dorsal region of the posterior column; *c*, tractus septo-marginalis.

The termination of part of the fibres of Gower's tract in the corpora quadrigemina region, without connection with the cerebellum, though stated by many authors, is not established with certainty.

Besides these long ascending tracts, there course in the lateral columns a great number of short fibres; they run upwards for a few segments, passing along in the remnants of the lateral column near the gray matter.

In the *anterior column* also, throughout the entire spinal cord, long bundles of fibres course upwards along the anterior fissure and the ventral periphery, to be gradually lost in its uppermost divisions. They come, in the

main, from the gray matter of the other side, from which they reach the anterior column through the anterior commissure. From the upper cervical portion of the cord, however, one can trace fibres, which, arising in the gray matter, reach the crossed anterior column by way of the anterior commissure, through the lateral division of the medial fillet and up to the optic thalamus, so that here a second spino-thalamic connection is present besides the above mentioned one of the long posterior tracts. Still other fibres from the anterior column reach the region of Deiter's nucleus by way of the reticular formation of the medulla oblongata. Besides these long paths, there are in the anterior column a large number of short ascending fibres, which after a few segments end again in the gray matter.

We can, therefore, group the conducting tracts of the spinal cord, as follows:

I. Descending tracts.

(*a*) *Lateral column.*

1. Lateral pyramidal tract (tractus cortico-spinalis cruciatus).
2. Rubro-spinal tract (Monakow's bundle).
3. Fibres from Deiter's nucleus.
4. Respiratory tract in the antero-lateral column.
5. Intraspinal fibres.

(*b*) *Anterior column.*

1. Anterior pyramidal tract (tractus cortico-spinalis ant.).
2. Vestibulo-spinal tract.
3. Corpora quadrigemina anterior tract.
4. Anterior pontile tract.
5. Respiratory tract.
6. Intraspinal fibres.

(*c*) *Posterior column.*

1. Descending posterior root fibres.
2. Descending endogenous fibres (Schultze's comma, Flechsig's oval field, medio-triangular tract).

II. Ascending paths.

(*a*) *Lateral column.*

1. Dorsolateral cerebellar tract (Flechsig) (tractus cerebello-spinalis dorsalis).
2. Ventrolateral cerebellar tract (Gowers) (tractus cerebello-spinalis ventralis).
3. Intraspinal fibres.

(*b*) *Anterior column.*

1. Ascending anterior column tract (faisceau sulco-marginal ascendant Marie).

2. Spino-thalamic tract from the upper cervical portion of the spinal cord.
3. Tract to the region of Deiter's nucleus.
4. Intraspinal fibres.

(*c*) *Posterior column.*

1. Ascending posterior column tract to the posterior column nuclei of the medulla oblongata: Goll's and Burdach's columns.
2. Posterior root fibres to the higher spinal cord segments.
3. Endogenous fibres (cornu-commissural zone and dorsal endogenous field).

Function of the Spinal Cord

If we now turn to the *function of the spinal cord*, we must first of all keep before our eyes the peculiar position of this central organ which, with the development of the brain, has sacrificed to it, more and more, the peculiar independence possessed by the cord in lower forms of animals and, as a whole, has become an intermediate organ between the centripetal stimuli coursing into the brain from the periphery of the body by way of the posterior roots, and the impulses sent from the brain centers to the peripheral organs. The former independent function of the spinal cord finds expression now only in *reflex activity*, and even this has in great measure come under the dominion of the brain centers. Only after the two have been completely separated, does a series of reflex phenomena appear in the isolated spinal cord, which must be attributed to its "isolation changes."

For the production of a reflex, there is needed, besides the afferent and efferent nerve paths, a connecting link, the *reflex center*. From the afferent nerve, the reflex center and the efferent nerve the *reflex arc* is formed, the functional intactness of which is necessary to the reflex action. For the spinal cord the *Magendie-Bell's law* is of fundamental importance; according to this only the anterior roots assume motor conductivity, whereas to the posterior alone is assigned the sensory conductivity, a law, which in its classic form, was first demonstrated in the frog by *Joh. Müller*. Even if in more recent times some observations have appeared, according to which, in the frog, some vasodilatation impulses leave the spinal cord by posterior roots, the general applicability of the law for mammals has not, as yet, been shaken.

By this law, the direction in which the reflex processes have to take place in the spinal cord, is firmly established. There are reflexes, which have their entire reflex arc in one segment of the spinal cord, whereas in other reflexes, various segments of the spinal cord, indeed even cerebral centers are concerned. In the entire conception of the reflex process it is

unquestionably a matter of importance, whether we must assume, according to the neuron theory, a leap of the centripetal stimulus over to the motor ganglion cells, or whether we must assume that the conduction of the stimulus takes place through a fibrillary network without spacial interruption. However that may be we, at any rate, will ascribe to the ganglion cells the main rôle in these reflex transitions, and will not consider the extracellular fibrillary net as the central place.

The well-known experiment of *Bethe* on the black-clawed crab, in which a separation of the afferent and efferent fibres of an antenna from the brain ganglion did not remove the possibility of reflex stimulation of the feeler, seems to speak for the possibility of the summation of the stimuli and the tonus without ganglion cells. But a simple transference of these relations to the reflex processes in vertebrates is not feasible.

For the production of reflexes, *Pflüger* has laid down a series of conduction laws, which we reproduce in abbreviated form (according to *Leyden* and *Goldscheider*):

1. *The Law of Equilateral Conduction for Unilateral Reflexes*

Whenever muscular movements strictly confined to one-half of the body follow as reflexes upon a stimulus, which affects a peripheral sensory nerve, these movements without exception and under all circumstances, are upon the same half of the body to which the stimulated sensory nerve belongs.

2. *The Law of the Symmetry of Reflexes*

If the change in the central organ, which is produced by a stimulated sensory fibre, has already evoked unilateral reflexes and, in its further spread, motors of the opposite half of the spinal cord are also excited, that is, bilateral reflexes are produced, always and under all circumstances only such motors are affected, as are already excited on the primarily affected side, so that bilateral reflexes are never produced in the opposite direction.

3. *The Unequal Development of the Reflexes on Both Sides of the Body in Bilateral Reflexes*

If the irritation of a sensory fibre induces reflexes in both halves of the body, and they are more intensive and violent on one side, than upon the other, the stronger are always upon that side, to which the stimulated centripetal fibre belongs.

4. *The Law of Intersensitive-motor Movement and Reflex-irradiation*

The direction from the sensory to the motor nerve in the central organ, is turned in the brain, from before backward, in the spinal cord from below upward, that is in both cases towards the medulla oblongata.

Even though these laws of *Pflüger* are on the whole valid, there are nevertheless exceptions. There are found exclusively crossed reflexes; even in the spinal cord reflexes may spread from above downwards, etc.

The movements executed by reflexes, have, on the whole, the character of purposeful reactions. As a rule there is a co-operation of several muscles. Many of these reflexes may be designated as *protective reflexes*, as, for instance, the decapitated frog's withdrawal of his foot from diluted acid, but even the reflexes that serve vegetative life, which regulate the activity of the bowels, the bladder, the act of birth, are quite suitable to the purpose.

Of the greatest importance for the entire spinal cord pathology was the discovery of the *patellar reflex* in 1875; this presents the most typical example of the *tendon reflexes*. These tendon reflexes consist in the contraction of a muscle, or of a group of muscles, on striking the tendon of the muscle. Even though *Westphal* at first would have it that these reactions are only an expression of the direct muscular irritation evoked by the shock, the view of *Erb*, that a spinal reflex is concerned, is the opinion, which at present is the generally accepted theory. In the patellar reflex the reflex arc is made up of the sensory branches of the crural nerve, the quadriceps center in the second to the fourth lumbar segment, the corresponding motor roots and motor fibres of the crural nerve. An injury in the realm of any of these divisions, and even in the quadriceps muscle itself, can abolish the patellar reflex, even though the reflex arc is not completely destroyed. This is proven by the return of the reflex in a cerebral disease which increases the excitability of the anterior horn cells even though it has been lost for a considerable period of time (hemiplegia in tabetics). In affections of the cerebro-spinal descending tracts, especially of the pyramidal tract, exaggeration of the patellar reflex to a high degree is always to be found. Even by irritation of the reflex center itself, such as is evoked by strychnine and tetanus poison, such a reflex exaggeration may be caused. In animals the complete separation of the spinal cord from the higher centers leads after a brief stage, during which the reflex is diminished, to a considerable increase of it. On the other hand, a series of pathological observations in man, in contrast to the animal experiments, seemed to prove a total loss of the tendon reflexes after complete transverse section of the spinal cord. But since in most recent times a series of trustworthy observations of the retention or even increase of the tendon reflexes with a totally divided spinal cord have been made, peculiar complications must be involved in those cases, in which the reflex is absent. Probably, in these cases, abnormal changes exist in the realm of the reflex arc itself.

Completely analogous to the patellar reflex is the *Achilles tendon reflex*, the center of which is situated in the upper sacral portion of the cord, and the *triceps-reflex* in the arm, the reflex center of which corresponds to the sixth to seventh cervical segment. In injury to the cerebro-spinal motor conduct-

ing tracts, there occurs not only an increase of the tendon reflexes, but there appear immediately in the legs peculiar clonic phenomena, the *ankle clonus* and the *patellar clonus*, which are induced by stimulation of the sensory nerves of the tendons and muscles, the tendons being in a constant state of tension.

In addition to the tendon reflexes, the *skin reflexes* are of great importance. In man the *plantar*, the *abdominal* and the *cremaster reflex* have attained clinical importance above all others. The reflex arc of the skin reflexes is not limited to the spinal cord, but reaches up to the brain, partly even up to the cortex of the cerebrum. The tactile reflex of the toes in the dog, which is elicited by touching the back of the foot, and is analogous to the human skin reflexes, is permanently lost after the removal of the center for the extremities in the cerebral cortex. In hemiplegia, the skin reflexes on the side of the paralysis are, at least temporarily, abolished.

Study of the tactile reflex of the dog in cases of disjunction of cerebrospinal tracts, has, as a matter of fact, proven that the construction of the reflex arc for the skin reflexes is very complicated. Not only the afferent limb, but also the efferent limb of the tactile reflex possesses two different conducting tracts (ascending posterior and anterior column tracts, descending pyramidal tract and rubro-spinal bundle), so that it can be made to disappear only after the destruction of either both afferent or both efferent tracts.

In most recent times, the *Babinski toe reflex* has finally proven, that at least one of the skin reflexes possesses, under pathological conditions, besides the cortical reflex arc, a subcortical one. The toes show under normal circumstances a distinct plantar flexion upon irritation of the sole of the foot; but in injuries of the brain or spinal cord, which, as a rule, lead to an increase of the tendon reflexes and to spasticity, a dorsal flexion of the toes appears. This phenomenon is the normal reflex process in children during the first year of life, and is only supplanted by plantar flexion, probably connected with the assumption of erect walking by man. Correspondingly, in the lower apes, such plantar flexion of the toes is lacking under normal conditions.

The plantar reflexes have their portals of entrance and exit in the first two sacral roots. The cremaster reflex, which consists in raising the testicles by contraction of the cremaster muscle, when the inner surface of the thigh is irritated, is carried by the first and second lumbar roots. The abdominal reflexes, which are not always constant, are divided into an upper, middle and lower; they are connected with the seventh to the eleventh dorsal roots.

Besides these skin and tendon reflexes, the spinal centers, that are connected with the sympathetic nervous system, govern a series of functions. The fibres of the spinal cord that pass to the sympathetic, begin only in

definite divisions, that is, from the first dorsal, occasionally, too, from the eighth cervical segment to the second or third lumbar segment, and from the second to the fourth sacral segment. The sympathetic fibres pass out from the spinal cord with the anterior roots, and reach the sympathetic ganglia (Fig. 51).

At the level of the first dorsal segment lies the *cilio-spinal center*, the irritation of which causes dilatation; its loss, contraction of the pupil. It is subject to the influence of a higher center in the medulla oblongata, so that a unilateral total division of the cervical spinal cord induces likewise miosis of the pupil on the same side; this, however, is transitory.

The sympathetic fibres from the dorsal and lumbar portions of the cord are of marked importance for the *innervation of the vessels.* Vasoconstrictors and vasodilators for the vessels of the skin, muscles, intestines, seem to be influenced by the spinal cord. Especially important are the centers, which, in the upper sacral portion, influence the *bladder, rectum, and the genital apparatus.* Experiments in recent years have proved, that even after disjunction of the gray substance of the lumbo-sacral portion, or after removal of the entire lower divisions of the spinal cord, the evacuation of the bowels and bladder is still regulated after a fashion, so that under pathological conditions, centers independent of the spinal cord step into active work; but it is certain, that the normally regulated function of bladder and rectum is under the control of the spinal cord; even where a total division of the cord occurs at a high level, there appears again, after a time, involuntary evacuation of the bladder, but at regular intervals. The centers for the detrusor and sphincter of the bladder and for the unstriped muscles of the rectum, lie in the third to fifth sacral segment, for the external sphincter ani in the fourth and fifth sacral segment, for the anal reflex in the fifth sacral segment.

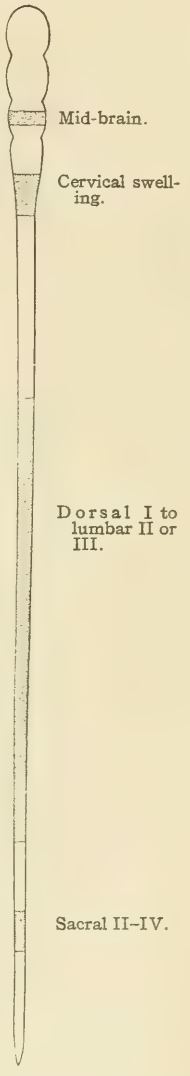


FIG. 51.—The autonomous nervous system in man. (*After Langley.*)

In the upper sacral portion of the cord, there are also the *centers* for *erection and ejaculation.* But here too the sympathetic ganglia, irrespective of the spinal-cord centers, seem to possess a certain independence of their own. Far more extensive is the same for the *female genital apparatus;* in animals with shortened spinal cords, as well as in human beings, after destruction of the lowest portion of the spinal cord, normal births have been observed.

The *independent function* of the spinal cord strikes us most clearly

when it is separated from the higher centers. In the decapitated frog, one can observe a great series of purposeful movements; thus, when one of its

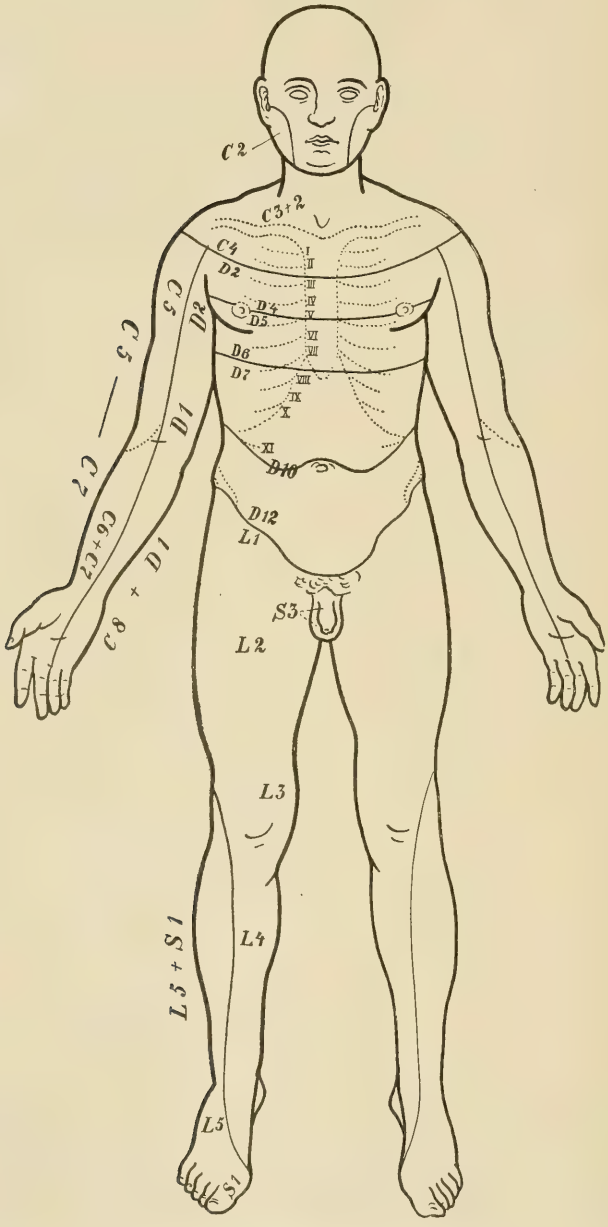


FIG. 52, *a*.—Scheme of cutaneous sensibility corresponding to the distribution of the posterior spinal roots. (*After Seiffer.*)

legs is held fast, it makes attempts to extricate it with the other, reacts with perfect appropriateness to abnormal skin stimulations, etc. But even in

the dog, peculiar reflexes appear after isolation of the spinal cord, as the well-studied scratch reflex, the rhythmical pawing of the hind legs, when the

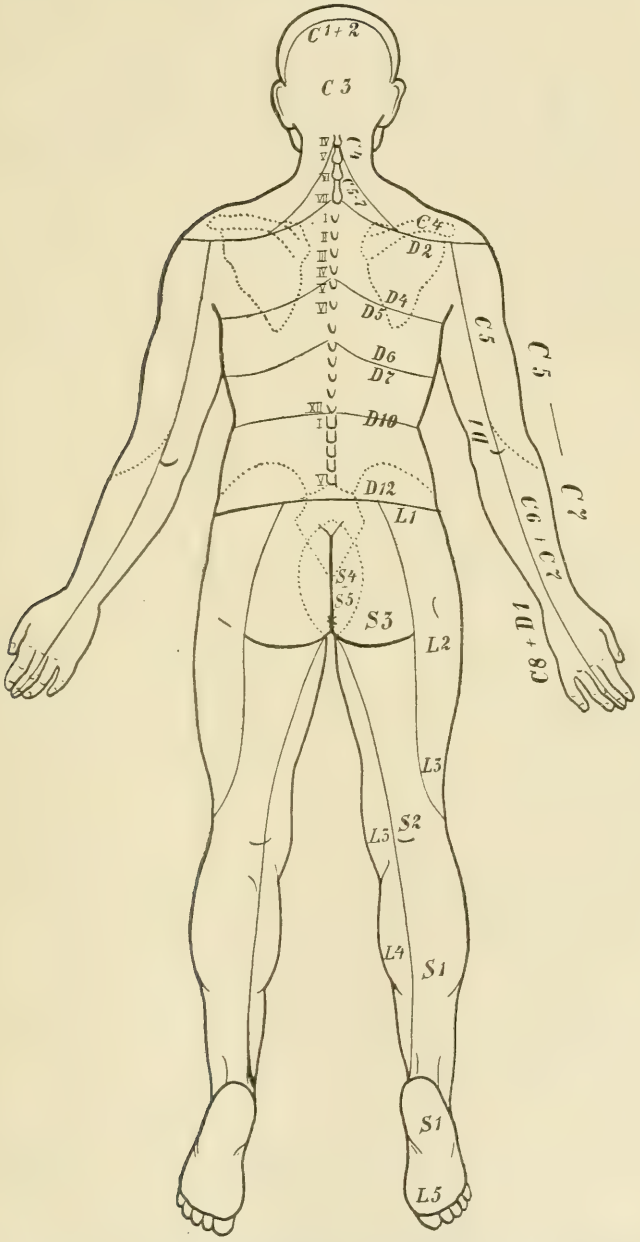


FIG. 52, *b*.—Scheme of cutaneous sensibility corresponding to the distribution of the posterior spinal roots. (*After Seiffer.*)

animal is lifted high up (Freusberg's phenomenon). Indeed, with forward motions of the front part of the body, which is connected with the brain,

there may be a lifting and a certain running-like movement of the hind legs, as the result of the reflex activity of the isolated spinal cord. In man, of course, such independence of the isolated spinal cord no longer exists. But here, too, there may be a series of abnormal reflexes and muscle twitchings.

While in many invertebrates the entire body is divided into numerous segments, to which a similarly divided central organ corresponds, this division in vertebrates is demonstrable only during embryonal life. In later life this *segmentation* is externally recognizable only in the vertebral column and in the origin of the spinal roots. In spite of this, every *spinal-cord segment*, characterized by the entrance and exit of definite roots, is in relation with definite parts of the surface of the body and definite skeletal muscles. Thus one may speak of a *segment innervation of the skin*, as every posterior root supplies a connected skin region. Animal experiments have taught us that the realms of the single posterior roots overlap, so that in order to produce total anæsthesia in a portion of skin, not only the main root but also the one above and below must always be destroyed. In addition to the schemes offered by these experimental investigations, schemes of the radicular innervation of the skin have been outlined relatively exact by the results of pathologic studies; first of all, by the phenomena of loss of function observed in tumors and compressions of the spinal cord, as well as by the peculiar trophic disturbances, corresponding to the sensory zones in herpes zoster, a disease based upon inflammation of individual spinal ganglia. In recent times they have come to be of the greatest practical importance for the exact localization of the height at which spinal-cord tumors exist and without which successful operative treatment is impossible.

The *sensibility scheme* (Fig. 52, *a* and *b*) of *Seiffer* reproduced here shows as the most important boundaries: the parietal-ear-chin line, which separates the trigeminus area from the cervical region; of next importance, the neck-trunk boundary, anteriorly it is in the second intercostal space, posteriorly it corresponds to the spines of the fifth to seventh cervical vertebræ where the zones of the fourth cervical segment and the second dorsal segment meet. After the fœtal outgrowth of the front extremities, the skin regions corresponding with the fifth cervical to the second dorsal segments, are distributed at the front and back of the arms. Of importance also are the intermamillary line (fourth to fifth dorsal segment), the xiphoid line (sixth to seventh dorsal segment), and the navel line (tenth dorsal segment). The trunk-leg line, formed in front by Poupart's ligament, behind by the first to second sacral vertebra, separates the twelfth dorsal and first lumbar segment. The divisions from the second lumbar segment to the second sacral segment serve for the skin innervation of the legs, while the lowest sacral segments subserve the genital and anal spheres. Thereby it is noteworthy that a lesion of the upper sacral portion may produce anæsthetic

disturbances at the inner edge of the foot and in the genital area, while the intermediate skin regions remain undisturbed.

But not only the skin innervation is dependent on the segmentary distribution of the roots, but in the *anterior roots* also there is a peculiar segmentary arrangement of the innervation of the muscles of the body, which swerves far from the distribution in the peripheral nerves and the muscles themselves. Thus every anterior root takes part in the innervation of several muscles, which may be supplied by different peripheral nerves, and again a series of anterior roots may take part in the innervation of one muscle.

The motor effects accomplished by the stimulation of anterior roots, have become known to us, as a whole, by experiments on monkeys. Concerning stimulation of anterior roots in man, we have only isolated information at our disposal (see following scheme).

The Motor Effects of the Electric Stimulation of the Anterior Roots in the Ape. (*From Sherrington.*)

Cervicalis.

I. Lateral flexion of the neck without rotation of the head.

II. Lateral flexion and retraction of the neck, very slight rotation of the head.

III. Lateral flexion with rotation and retraction of the neck, so that the chin is correspondingly turned towards the opposite side.

IV. Elevation and adduction of the shoulder. Retroflexion and slight lateral flexion of the neck, by which the head, especially when the shoulder is "fixed," is drawn to the contra-lateral side.

V. Elevation, abduction and slight outward rotation of the shoulder. Flexion of the elbow, at the same time slight supination and radial flexion of the hand. Only slight lateral and retroflexion of the neck.

VI. Moderate adduction of the shoulder. Strong flexion of the elbow. Slight extension of the fingers and the hand, but in some individuals flexion. Some supination. Neck and head as in V.

VII. Retraction and pronounced adduction of the shoulder and inward rotation of the upper arm. Extension of the lower arm. Slight flexion and pronation of the hand.
Slight flexion of the fingers. The shoulder is drawn down. Neck as in V.

VIII. The shoulder is drawn down (latissimus dorsi). The adduction of the shoulder is not so strong as in VII. Inward rotation

of the arms. Flexion and pronation of the hand. Flexion of the fingers and the thumb with apposition of the latter.

Thoracica.

I. Retraction of the shoulder. Slight lateral and retro-flexion of the neck. Slight extension of the arm. Flexion and pronation of the hand. Flexion of the fingers and the thumb with opposition of the latter. Usually, also, slight ulnar flexion of the hand.

II. Retraction of the shoulder. Slight flexion of the hand. Flexion of the fingers and the thumb with opposition of the latter. In some cases, slight pronation of the hand. Lateral flexion of the vertebral column.

Postthoracica (lumbalis).

I. Drawing in of the abdomen.

II. The same with slight flexion of the hip.

III. Drawing in of the lower part of the abdomen. Flexion of the hip.

IV. Drawing in of the lower part of the abdomen. Flexion and adduction of the hip. Extension of the knee.

V. Adduction of the hip. Extension of the knee. Slight flexion of the foot; slight extension of the great toe.

VI. Extension of the hip. Adduction of the thigh. Strong flexion of the knee. Dorsal flexion of the foot. Extension of the toes. Adduction of the hallux.

VII. Extension of the hip. Flexion in the knee. Extension of the foot. Rotation of the sole. Strong flexion and adduction of the hallux. Downward movement of the tail.

VIII. Lateral movement of the tail. Slight outward rotation of the hip and flexion in the knee, with extension in the hip and foot. Strong flexion of the toes with flexion and adduction of the hallux.

IX. Lateral movement of the tail. Sometimes, slight outward rotation of the thigh and adduction with flexion of the toes.

X. Lateral movement of the tail (towards the side of stimulation).

These discoveries prove so much, that the assumption of a definite functional relation of the motor fibres united in one anterior root, is not justified. Furthermore, it is noteworthy that even for paralysis of one muscle, at least three successive anterior roots must as a rule be destroyed. In man, frequently, a still farther reaching anastomosis of the individual roots is assumed, so that beside the middle root, two upper and two lower roots take part in the innervation of a muscle.

Observers have been largely occupied, in ascertaining the localization of the various muscles in the individual spinal-cord segments, from the results of localized spinal-cord lesions. From the wealth of schemes put together for this purpose, we give the one recently published by Bruns, as it takes into consideration in addition skin innervation and the reflexes.

LOCALIZATION OF THE FUNCTIONS OF THE VARIOUS SEGMENTS OF THE SPINAL CORD. (*From Bruns*)

Segments	Muscles	Sensory innervation of the skin	Reflexes
1. Cervicalis.	Musculus rectus capitis posterior minor.[1]		
	M. rectus capitis posterior major.		
	M. obliquus cap. superior.		
	M. semispinalis capitis.		
	M. spinalis cap. (pars cranialis).		
	M. rectus cap. anterior.		
	M. longus capitis.		
	M. rectus capitis lateralis.		
	M. geniohyoideus.		
	M. omohyoideus.		
	M. sternohyoideus.		
	M. thyreo-hyoideus.		
	M. sternothyreoideus.		
	One branch to the musculus intertransversarius posterior cervicalis.		
2. Cervicalis.	M. rectus cap. post. major.		
	M. obliquus cap. inferior.		
	M. semispinalis capitis.		
	M. spinalis capitis (pars cranialis).		
	M. longus atlantis.		
	M. longus colli.		
	M. longus capitis.		
	M. geniohyoideus.		
	M. omohyoideus.		
	M. sternohyoideus.		
	M. thyreohyoideus.		
	M. sternothyreoideus.		

[1] Cervicales, 1 to 4, from Risien Russel.

Segments	Muscles	Sensory innervation of the skin	Reflexes
	M. splenius capitis et cervicis. M. sternocleidomastoideus.[1] M. trapezius. Serrations of the musculus longissimus and the intertransversarii.	Skin on the back part of the head to the vertex; on the sides of the head forwards to the anterior boundary of the ear; skin over the ear; over the lowest and most posterior parts of the lower jaw, on the neck and nape, at the front downwards to the second rib; at the back down to the spina scapulæ (according to Wichmann, the middle lower part of this dorsal region is supplied by the dorsal branches of the fifth and sixth and even seventh cervical nerves). The second cervical root reaches upwards directly to the trigeminus region; the third innervates the neck and nape down to the clavicle; the fourth from there to the second rib.	
3. Cervicalis.	Platysma (Kocher). M. longus atlantis. M. longus colli. M. longus capitis. M. diaphragma. M. scalenus medius. M. geniohyoideus. M. omohyoideus. M. sternohyoideus. M. sternothyreoideus. M. splenius capitis et cervicis. M. sternocleidomastoideus. M. trapezius. M. levator scapulæ.		
4. Cervicalis.	M. longus atlantis. M. longus colli. M. longus capitis. *M. Diaphragma.* M. scalenus anterior. M. scalenus medius. M. sternothyreoideus. M. splenius capitis et cervicis. M. trapezius. M. levator scapulæ. M. rhomboidei. M. supra- and infra-spinatus. M. deltoideus. M. biceps and coracobrachialis. M. brachialis int. M. supinator longus.		

[1] Through accessorii.

Segments	Muscles	Sensory innervation of the skin	Reflexes
5. Cervicalis.	Musculus levator scapulæ.[1] M. teres minor. M. diaphragma. M. rhomboidei. M. deltoideus. M. biceps and coracobrachialis. M. brachialis internus. M. supra- and infra-spinatus. M. supinator longus and brevis. M. pectoralis (pars clavicularis). Possibly extensors of hand. M. serratus anticus major. M. splenius.	Outer side of shoulder, arm and forearm.	Scapular reflex. From the fifth cervical to the first dorsal root, tendon reflexes of the muscles represented in them.
6. Cervicalis.	M. deltoideus. M. teres major and minor. M. biceps and brachialis internus. M. supra- and infra-spinatus. M. supinator longus and brevis. M. subscapularis. M. pronator quadratus and teres. *Extensors and flexors of the hand.* Serratus anticus major. Possibly long extensors of the fingers. M. splenius. M. scaleni.	Outer parts of the forearm on the flexor and extensor surfaces. Radial portion of the hand and the fingers.	
7. Cervicalis.	Pronators of the hand. M. subscapularis. M. teres major. Extensors and flexors of the hand. *M. triceps.* Pectoralis major (pars costalis). Latissimus dorsi.	Middle parts of the forearm on the flexor and extensor surfaces. Radial and median district of the hand and the fingers.	Palmar reflex. Blow upon the palmar surface causes the fingers to close.

[1] Besides this, the individual cervical roots innervate also the long muscles, extending over the entire vertebral column, at the corresponding level.

Segments	Muscles	Sensory innervation of the skin	Reflexes
	Long extensors and flexors of the fingers. M. splenius. M. scaleni. Small muscles of the hand (interossei and lumbricales)?		
8. Cervicalis.	M. triceps. Flexor carpi ulnaris. Long extensors and flexors of the hand. *Interossei and lumbricales.* Scaleni.	Ulnar side of the hand, the forearm and arm on the extensor and flexor surfaces. The uppermost part of the arm region is supplied in addition by dorsalis 2.	
1. Dorsalis.	Long flexors of the fingers. Interossei and lumbricales. Balls of the *thumb and small finger.* Scaleni. (In the eighth cervical and first dorsal root the fibres for the dilatator pupillæ pass to the gangliated cord.)		
2 to 12 dorsalis, resp. 1 lumbalis.	The corresponding long muscles of the vertebral column. M. serratus post. sup. 1–4. M. serratus post. inf. 9–12. M. intercostales interni 2–11. M. intercostales externi 2–11. M. levat. cost. breves 2–11. M. infracostales 2–4 and 7–9. M. levat. cost. longi 8–10. M. transversus thoracis 3–6. M. transversus abdominis 7–12.	Skin of the chest, the back, and the abdomen, above from the lower boundary of the upper cervical area, below reaching not quite to the inguinal fold; skin of the upper gluteal region. The 6th root reaches in front, medially, the region of the epigastrium; the 8th and 9th do not reach beyond the skin over the abdominal cavity (nipple 4th dorsal root; upper epigastrium 6th, umbilicus 10th). The boundaries of the individual skin regions pass horizontally about the trunk; therefore, at the thorax, over several intercostal spaces.	Epigastric reflex, 4th–7th dorsal root. Abdominal reflex, 7th–11th dorsal root.

Segments	Muscles	Sensory innervation of the skin	Reflexes
	M. obliq. abd. int. 7–12. M. obliq. abd. ext. 7–12. M. rectus abdominis 8–12. M. pyramidalis d. 12 lumb. 1.		
Lumbalis 1.[1]	Muscles of the abdomen, see above. Iliopsoas. Sartorius. Cremaster. Quadratus lumborum.	Skin of the inguinal region, of the uppermost part of the front and outer parts of the thigh, skin of the mons veneris, the root of the penis on the dorsal side, sensibility of the testicles, seminal cord and the tunica dartos, by means of the sympathetic.	Patellar reflex 2nd–4th lumbaris, especially lumb., 4.
Lumbalis 2.[1] *Lumbal plexus.*	Iliopsoas. Sartorius. Quadriceps femoris. Cremaster. Quadratus lumborum.	Outer and front part of the hip, to the lower third of the thigh.	
Lumbalis 3.	Small muscles of the pelvis. M. iliopsoas. M. sartorius. M. quadriceps femoris. M. adductores femoris. M. quadratus lumborum.	Anterior portion of the hip; partly in common with 2, innermost parts of the back of the hip in its upper parts, and the inner side of the leg.	
Lumbalis 4.	Small muscles of the pelvis. M. quadriceps femoris. M. adductores femoris. Tibialis anticus. (Extensor digitorum commun., extensor hallucis.)	Lower parts of the inner and anterior surfaces of the hip, inner side of the leg, partly also the back inner margin of the foot, excepting the large toe.	Glutæal reflex 4th and 5th lumbalis (? probably lower).
Lumbalis 5.	Tibialis anticus. Extensor hallucis. Extensor digitorum communis. Musculi peronei.	Anterior outer parts of the leg, outer and dorsal parts of the foot, with exception of the tips of the toes, inner parts of the plantar surface of the foot (with sacralis).	

[1] Perhaps the 1st and 2nd lumbar segments contain no muscular nuclei at all; then the muscles mentioned here would draw their nerve supply from the 3d segment.

Segments	Muscles	Sensory innervation of the skin	Reflexes
Sacral plexus.	M. flexores cruris.[1] Musculi glutæi (extensores) and abductors of the hip. Outward rotators of the hip. Plantar flexors of the foot (musculature of the calf) and long flexors of the toes.		
Sacralis 1 and 2.	M. peronei. Musc. flexoris cruris. Muscul. glutæi (extensors) and abductors of hip. Outward rotators of the hip. Plantar flexors of the foot. Long flexors of the toes.	First sacralis. Outer and middle part of the sole, heel, parts of the dorsum of the foot (inner side), tips of the toes. Outer and back part of the calf.	Plantar reflex.
	Second especially: small foot muscles, interossei and flexor digitorum brevis.	Second sacralis. Middle part of the back of the thigh and leg.	Achilles tendon reflex. According to Ziehen, sacralis 1; according to Oppenheim, lumbalis 5, sacralis 1 and 2.
Sacral plexus. Sacralis 3–5.	Muscles of the perineum and the transversely striated musculature of the urethra (compressor urethræ) and of the rectum (sphincter ani externus) and of the sexual organs.	Third sacralis. Skin over the os sacrum, inner and upper part of the thigh.	Center for erection 2 and 3 sacralis. For ejaculation 3 sacralis.[2]
		Fourth and fifth sacral root. Perineum, anus, and concentric parts of the skin over the os sacrum close to the middle line, dorsum of penis, and anterior surface with the exception of the root of the penis, skin of the scrotum.	Detrusor and sphincter vesicæ, unstriped musculature of the rectum 3, 4, and 5 sacralis.[2] Sphincter ani externus 4 and 5 sacralis.
		(In the female, back parts of the large labia.)	Anal reflex 5 sacralis.

[1] The segments for the flexores cruris and the glutæi are not yet clearly determined. The former lie certainly higher than the latter; they may even be below the muscles of the calf. Both lie certainly below the peronei.

[2] According to L. R. Muller, not situated in the conus.

In recent times, with the aid afforded by the *methods of chromatolysis* in amputations and muscle atrophies, it has been attempted to establish more exactly the *relations of the individual columns of nuclei in the anterior horns to the muscle groups of the extremities.* According to these investigations, for instance, every cell column of the lumbar enlargement seems to stand in a certain relation to all the muscles of a limb segment. Each of these segmentary groups of nuclei might be divided into individual nuclei, which are connected either with the function of similar groups of muscles, or with single nerves of the limb segment concerned. Finally, there might be subdivisions here again for individual muscles. With these investigations as a basis, schemes of the various columns of nuclei in the cervical and lumbar enlargements with their proper functions have already been drawn up, though no decisive results could be gained here.

If we until now have mainly been treating of the functions localized in the individual spinal cord segments, we must now turn to the *functional importance of the conducting tracts* of the spinal cord. Their anatomical distribution we have discussed above.

The investigation of the *function of the spinal cord tracts* has undergone great changes with the advancing knowledge of their anatomical structure. Only since we have been precisely informed as to the course of the individual tracts, which are often mixed in the same area, has it become possible to confine their experimental disjunction to one or a few of them. Besides, the accurate microscopic observation of the point of incision and the secondary degenerations, in which Marchi's method has been of the greatest assistance, places in our hands the indispensable means of controlling exactly the prearranged injuries as to extent, as well as the manifestations that follow.

In addition to the experimental investigations in higher mammals, the experience afforded by human pathology has been of great importance. In very truth, it is indispensable for the ascertainment of the function of the conducting tracts in man, since not only in anatomic structure, but also in the physiological value of the individual tracts, there are important differences between the various classes of animals and man. But one must always remember, that in the sense of physiological experiment pure disjunctions of definite divisions of the spinal cord in human pathology are the rarest of exceptions, since pressure phenomena, focal diseases due to other causes, etc., are quite liable to confuse the pure symptoms of lost function.

Considering the gradual development of our knowledge of the cerebrospinal conducting tracts, it is easy to understand, how observers were immediately inclined to assign the individual functions to the few conducting tracts that were known. Thus the *pyramidal tract,* especially, was long considered as the only pathway for transmitting voluntary motor impulses from the cortex of the cerebrum to the spinal cord. This conception found its actual support in the proof of descending degeneration of the pyramidal

tract in man after focal injuries to the brain, especially to the inner capsule in connection with a typical hemiplegia. But it must be borne in mind, that in this case, though only the pyramidal tract degenerates down to the spinal cord, it was not alone interrupted in the brain by the focal injury, but in connection with the other cortico-fugal tracts subserving the region of the extremities. But even these wide spread disjunctions of the tracts, that carry the motor impulses, do not prevent a considerable involution of the hemiplegia after some time.

An *isolated disjunction of the pyramidal tract* is experimentally feasible only by complete division of the pyramids in the medulla oblongata, where they appear on the ventral surface and are not mixed with other fibres, or by section of the pyramidal crossing itself making a longitudinal incision, which at least produces, in the dog and lower ape, complete disjunction of the pyramidal conductivity, since in them the anterior pyramidal tract is wanting. In the lateral column of the spinal cord, however, the pyramidal tract can be severed only in conjunction with the rubro-spinal and dorso-lateral cerebellar tract.

While it had been known for some time, that after severing the pyramidal tracts the motor effects in electric stimulation of the cortex of the cerebrum remain unchanged, it has recently been possible to keep dogs alive for a long time after the pyramidal tracts had been completely cut through. Thus it was seen that the animals, immediately after waking from the narcosis after the operation, were able to run about freely and, in the use of their extremities, could not be differentiated from normal dogs. In unilateral disjunction of the pyramids, a certain awkwardness in the employment of the muscles of the opposite side of the body was noted in performing unusual movements (waddling); but nothing of functional loss was observed. Correspondingly, the isolated disjunction of the pyramidal tracts does not essentially injure either the tactile reflex, passing over the region of the extremities of the cerebral cortex, nor the effects of electric stimulation of the region of the extremities.

Now, if the second important motor tract in the lateral column of the dog is disjoined, the *rubro-spinal bundle* in the medulla oblongata above the pyramidal crossing, that is, before the pyramidal fibres enter the lateral column, there appears immediately clearly defined paresis of the limbs on the same side, which, however, is completely recovered from in the next few weeks. In this, also, electric stimulation of the region of the extremities shows no deviation from the normal; the tactile reflex, lost at first, is soon recovered again (Fig. 53).

Not until a *disjunction of both the pyramidal and the rubro-spinal tracts* is performed in the lateral column of the upper cervical portion of the cord of the dog, are serious phenomena of loss of function observed. After a temporary, rather serious paralysis of the extremities of the same side, a spastic

paresis of the limbs remains. The tactile reflex is permanently lost; isolated movements of the extremities are no longer observed. Above all, the effect of the electric stimulation of the crossed region of the extremities is absolutely lost. In the dog, therefore, the impulses of the cerebral cortex are transmitted to the spinal cord exclusively by way of the direct cortico-spinal (pyramidal) and the rubro-spinal tracts.

In the *lower apes* also the *total destruction of the pyramidal crossing* has the surprising result, that no paralysis of the extremities appears, not even immediately after the operation. But not only the coarser voluntary movements, as they appear in the acts of running and climbing, are retained, but also the delicate isolated movements of the fingers which are necessary, for instance, in reaching for small pieces of food, are absolutely intact. On the whole, however, the ape, in whom the pyramid has been cut, performs all his movements more slowly than the normal ape, and thus makes altogether a clumsier impression (Fig. 54).

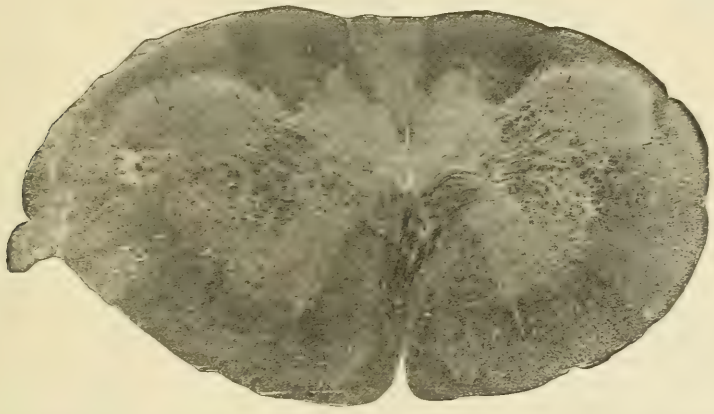


FIG. 53.—Division of the right postero-lateral column including the rubro-spinal bundle at the height of the pyramidal crossing in the dog. Marchi preparation.

But that the pyramidal tracts are of considerably greater importance in the ape than in the dog, is proved by the result of *cerebral cortex stimulation;* when this is performed several weeks after the disjunction of the pyramidal tracts, only two small fields of the region of the extremities, those corresponding to the centers for finger and toe movements, are proven electrically excitable, whereas all the rest of the field does not respond.

Nor does the isolated disjunction of the *rubro-spinal tract of the lateral column*, before it joins with the pyramidal tract, lead to any paresis, worth mentioning, of the extremities on the same side. In unilateral disjunction they are somewhat more awkward for a few days than the extremities of the other side. But after a few days they are used quite normally for all purposes, including the most delicate grasping movements, with the exception that the normal arm is somewhat preferred. To this corresponds also

the completely normal electric excitability of the crossed region of the extremities in the cerebral cortex.

Now, if in the ape, both pyramidal and rubro-spinal tracts are interrupted together, by complete division of the postero-lateral column in the third cervical segment, the result differs materially from that obtained in dogs. On the first two days after such an operation the extremities concerned hang down limply, and are but slightly used in the general movements. But in the next few days, the motor power of the extremities gradually increases; arms and legs are used not only in general, but possibly even better in isolated movements. Indeed, after a week the motor weakness is virtually wholly adjusted. After special practice (the sound arm is bound down for some

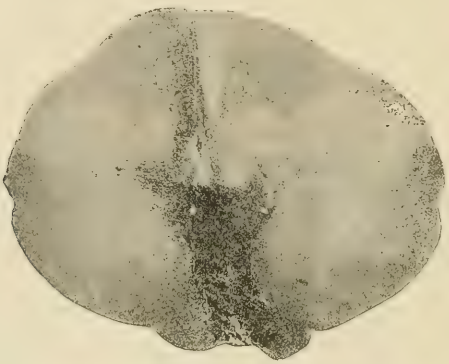


FIG. 54.—Division of the pyramidal crossing in the ape (Macacus rhesus). Marchi preparation.

time every day) the ape prefers this arm, even for grasping. In the ape, in contradistinction to the dog, and still sharper contrast to man, no spastic phenomena follow such lateral column disjunction.

It is in full accord with this result, that in the ape the electrical stimulation of the region of the extremities several weeks after a unilateral disjunction in the postero-lateral column still transmits motor impulses to the spinal cord, which, though they are virtually confined to the fields of the hand and foot region, are still somewhat more marked than may be observed in bilateral isolated pyramidal division.

Since, then, the combined disjunction of the cortico-spinal and the rubro-spinal tracts in the ape leads only to a slightly greater disturbance of the voluntary movements of the extremities, than the isolated disjunction of each of these tracts, other tracts must be involved in their conduction to the spinal-cord centers. In the first place it can easily be proven that it is not only, as one might think, the corresponding conducting tracts of the other half of the spinal cord which permit the impulses to cross over to the injured side at the level of the corresponding anterior-horn centers. Even in the dog, *bilateral disjunction of the postero-lateral column* leads to no serious disturbance so far as the usual locomotion is concerned. In apes,

also, to the complete division of one postero-lateral column, a complete division of the other may be added after 2 to 3 weeks, and still the ape can learn to run and climb again after a few days, and is able, though with lessened vigor, to reach for food. Furthermore, even if in the ape first the entire arm region of one hemisphere of the cerebrum is removed to the extent marked out by *Munk*, and the isolated grasping movements of the opposite arm are thereby lost by complete destruction of the corresponding pyramidal tract, and to this is added, after a few weeks, the disjunction of the postero-lateral column of the same side in the third cervical segment, which has been innervating the heretofore normal extremities, he can nevertheless soon learn to use these extremities again for general movements and isolated grasping motions. Indeed, the disjunction of the impulses of the other side can be carried still farther, by cutting through the same ape's other postero-lateral column in the first cervical segment, thus disjoining the one remaining rubro-spinal bundle and the few uncrossed pyramidal fibres which radiate into this column, only to find that the restitution of the arm, which still has its cerebral cortical centers, is not impeded, though the motor lateral column tracts on both sides are totally disjoined. These movements of the extremities concerned are present even though only at first to a very limited extent, immediately after complete division of the postero-lateral columns, so that an absolute restitution should not be spoken of, as part of the motor impulses must already be passing normally outside of the motor lateral column tracts of the spinal cord (Fig. 55).

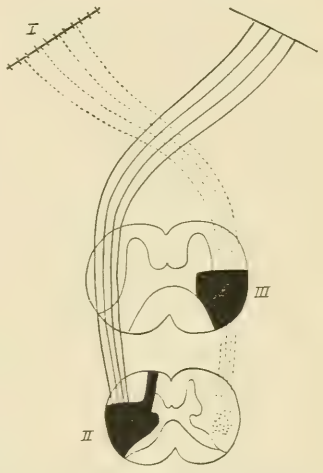


FIG. 55.—Combined disjunction of one arm region and both postero-lateral columns. *I*, Extirpation of the left arm region (*After Munk*); *II*, complete division of the left postero-lateral column and of parts of the anterior column in the third cervical segment; *III*, complete division of the right postero-lateral column in the first cervical segment.

Here, then, the *anterior columns* must be considered; in them we have already met with (see above) a series of cerebro-spinal tracts passing to the spinal cord. Older physiologists already ascribed to them the most important share in the conduction of movements. But, nevertheless, they were later neglected as compared with the pyramidal tract, inasmuch as even human pathology furthered the knowledge of the motor functions of the anterior columns but slightly. *Isolated disjunction of the anterior columns* may be performed in dogs, from the front, along the lower margin of the atlas, directly beneath the pyramidal crossing. Such an isolated disjunction of the anterior columns leads to no paralysis of the extremities. On the other hand, the anterior columns subserve a function especially important for the

station of the animal, namely, the innervation of the musculature of the back. The dog, deprived of his anterior columns, runs in a wobbly fashion, with legs apart and with the back part of his body shaking from side to side. He can not turn sideways in a normal way, and when he is put on the table with the back part of his body hanging down over the edge, he can not raise it as a normal dog can. Electric cerebral cortex irritation of the regions of the extremities, however, does not show the slightest loss of function, after the anterior column has been disjoined.

In the dog, therefore, neither the disjunction of the lateral, nor that of the anterior columns, brings about paralysis of the extremities, though under normal conditions the motor impulses, which leave the cerebrum, according to the results of stimulation of the cerebral cortex, seem, on the whole, to use the lateral column. Since, on the other hand, the disjunction of the pyramidal tract together with the anterior column, stops neither the motor activity of the extremities, nor the effect of stimulation of the cerebral cortex, we reach the important conclusion, for the dog at least, that *all these motor lateral column and anterior column tracts can mutually, to a great extent, replace each other*, with the exception that the impulses for the isolated movements, dependent exclusively on the cortex of the cerebrum, use the lateral column tracts par preference.

In the monkey, an *isolated disjunction of the anterior column* has not yet been attempted. It has, however, been ascertained, that in the ape also a destruction of the anterior column together with the pyramidal tract does not cause any loss of motor function of the extremities concerned, nor a total abolition of the electric cortical excitability, so that the rubro-spinal tract alone, might, even in the ape, suffice to conduct these impulses. But there can be no doubt, according to the above quoted results of disjunction of the lateral column, that the *anterior column* possesses in the *ape* a higher physiological value than in the dog, since it is used in great measure also for the conduction of the isolated movements dependent on the cerebral cortex, even to the most delicate play of the fingers.

But now, if in the dog or monkey the *entire motor conduction of half the spinal cord* is disjoined, either by induction of a complete hemilesion, or by severing only the anterior and lateral columns together in the upper cervical portion, it appears that even then motor impulses from the other half of the spinal cord may reach the spinal centers of the extremities concerned. Thus, after transient serious paralysis a moderate degree of movement is regained, even though the excitability of the opposite cerebral hemisphere is lost, and in the monkey, at least, the grasping movement of the affected arm remains seriously injured.

The remarkable power of restitution which, with respect to coarse locomotion, is innate in the spinal cord, is best demonstrated by an experiment on the cat, in which the motor tracts of the lateral column, the medial

halves of the anterior columns, and in addition the posterior columns on both sides, were destroyed. In spite of the fact that the cat was totally paralyzed in all four extremities for 2 1/2 weeks, ordinary walking movements gradually returned, which improved to such an extent in the course of 1 1/2 months, that the cat was able to reach by jumping, and to bring down meat from the edge of a table of ordinary height. For this action, which certainly demands a considerable amount of strength and agility, the lateral remnants of the anterior columns sufficed in this case, though the extensive destruction of sensory fibres had certainly increased the difficulty of restitution.

In the *anthropoid ape*, experiments concerning the functions of the motor conducting tracts, which, as is easily understood, would be of especial value particularly because of the far reaching analogy of its anatomic structure with that of man, have been carried out only in isolated cases. The destruction of the smaller medial half of one pyramid immediately above the pyramidal crossing, is, in the chimpanzee, followed by slight weakness in the arm concerned, without decided phenomena of paralysis, without any indication of spasms, and with retention of the delicate grasping movements of the fingers. A destruction of the medial half of one anterior column, including the anterior pyramidal tract, which in the chimpanzee already appears in the second cervical segment, caused, likewise, but a slight tendency to spare the extremities on the same side, especially the arm, without the least disturbance even of the most delicate finger movements.

Concerning the functions of the *motor spinal cord tracts in man*, it was supposed, up to most recent times, that the *pyramidal tract* represented the path of voluntary movements and that its destruction was necessarily accompanied by paralysis of the corresponding extremities, with spasms and exaggeration of the tendon reflexes. If in hemiplegias, the result of lesions in the internal capsule, a return of voluntary movement could, as a rule, be noted after a shorter or longer period of time, this was supposed to be due to the fact that part of the pyramidal fibres were preserved. But after the possibility of almost complete functional replacement of the pyramidal tracts had been proved by experiments on animals, especially by those on such monkeys, which in the structure of their nervous system and in the functions of their extremities, especially the arms, resembles man, a revision of the former theory in man was also unavoidable. The exact study of cerebral hemiplegias proved, that in spite of a totally destroyed pyramidal tract, an extensive restitution of the motor function might occur, which, if the defects be acquired in earliest childhood, may even approach the normal conditions. Even in spinal hemiplegia, due to total destruction of one lateral column in the spinal cord, there is a restitution of approximately the same extent. In bilateral loss of the lateral pyramidal tract, as it has been observed in approximately pure form in some cases of "spastic spinal pa-

ralysis," there is no serious paralysis, but only a serious hindrance in walking because of rather severe spasms in the legs.

As all these results prove with certainty that *the loss of the pyramidal tracts is not followed, in man, by an irreparable loss of voluntary movements*, there remain only as consequences of the loss of the pyramidal tracts, exaggerated tendon reflexes with clonus, as well as the Babinski toe reflex and spasms. Whether the latter are an absolutely necessary result of the loss of pyramidal functions is, to say the least, questionable. In human pathology cases without injury of the pyramidal tracts have been observed with severe spasms, as well as other cases in which diseases of the pyramidal tracts were present with scarcely a suggestion of spasms. But it must be granted, that the combination of disease of the pyramidal tract and muscle tonus increased to spasm represents the rule.

But even though the pyramidal tract is unquestionably capable of a considerable measure of replacement in man too, still its importance is far greater, than it is in all animals up to apes, and it seems practically certain, that *sudden isolated interruption in the conductivity of the pyramidal tracts*, as occurs in rare cases in the medulla oblongata, is *immediately followed by serious paresis.*

Considering now the question of *compensation tracts for the pyramidal tracts* in man, *the rubro-spinal bundle* seems to have become so rudimentary here, that it can scarcely be considered in the restitution of the motor function. So far as the *anterior pyramidal tract* is concerned, which, of course, escapes in spinal diseases of the lateral column, and might possibly be of importance for motor conductivity, nothing certain is yet known about its function. We know of no difference between the symptoms of total destruction of the pyramidal tract above the crossing, and a lesion of the lateral column, with the anterior pyramidal tract intact. The above mentioned experiment, in which the anterior pyramidal tract of one side, was disjoined in the chimpanzee, seems to make it apparent, that it is not connected in any way with the other half of the spinal cord. This leaves the other *anterior motor tracts* to compensate for the pyramidal tract, analogous to the well established relations in the lower ape. Furthermore, pathology proves definitely, that in extensive destruction of the anterior tracts, the conductivity through the lateral pyramidal tracts suffices for the retention of motility.

And finally in man also, after destruction of *one entire half of the spinal cord*, the motor conductivity of the other side may bring about considerable recovery, especially for the leg of the injured side. In spite of initial serious paralysis of the leg, such individuals can, months and years afterwards, take long walks without assistance. Insufficient observations for satisfactory conclusions relative to this matter have been made concerning the arm.

Since, therefore, the pyramidal tract can be replaced almost completely in higher mammals, and very largely in man, by means of the other motor spinal cord tracts, so that it is not possible to ascribe to it a specific function, this question arises: of what importance is the greater and greater development, of such an uninterrupted cortico-spinal tract in the ascending animal series? The supposition, that this direct connection between the cerebral cortex and the spinal cord is necessary for the individual's learning of special motions, is contradicted by the established fact, that the ape, even after being deprived of his pyramidal tracts, can be taught new and complicated arm movements. Since the cerebral cortex can influence the spinal centers through the pyramidal tracts only without the assistance of the subcortical brain centers, it is probable, on the contrary, that in the learning of new movements, the influence of these latter centers upon the gray matter of the spinal cord is necessary for clearing the path of the groups of ganglion cells in the anterior horns, which are essential to this. But when the movement is completely controlled, the direct transference of the impulse from the cerebral cortex to the spinal cord, with its anterior horn cells already arranged for the definite combinations of muscles, suffices and the centers of the mid- and hind-brains may be passed over. *The pyramidal tract is, accordingly, the real conducting tract for the immense number of movements already acquired;* the cerebro-spinal tracts, on the other hand, that are interrupted in the mid-brain, with their relations to the cerebellum, are of the utmost importance in learning movements.

In turning now, to the *function of the ascending tracts* of the spinal cord, we shall first consider the disturbances, which are caused by *complete elision of all the sensory impulses of an extremity passing through the posterior roots to the spinal cord.* For instance, if, in the ape, we cut away all the posterior roots concerned with the arm, from the fourth cervical to the fourth dorsal segments, there is not only, as is to be expected, total loss of all sensory conductivity; but the grasping movements of the arm concerned also disappear at first. It was therefore supposed, that through this injury the isolated movements dependent on the region of the extremities in the cortex were being seriously impaired, or even destroyed with rather slight injury to the general movements, thereby proving the necessity of the impulses carried from the skin and muscles to the central organ for the execution of the most highly organized movements. But, in contradistinction to this, the most recent investigations have shown, that in fact, all normal movements of the extremities are injured by the loss of all the centripetal impulses. If one avoids all collateral injuries of the spinal cord, the disturbances of the general movements, which are dependent on the subcortical centers, remain unchanged. The grasping movements, however, which are principally or entirely dependent on the centers of the cerebral cortex, improve under the influence of practice; the loss of the stimulations, that normally pass to the

region of the extremities through the sensory tracts, is extensively compensated for by the other sensory impulses, especially those of the visual sense.

Concerning the *function of the individual ascending conducting tracts of the spinal cord*, the conducting paths for the *various forms of sensation* must be regarded separately, whether one assumes now that they are specifically differentiated from the periphery, or that they acquire their peculiar characteristics only with the aid of the spinal centers. We must therefore consider separately:

1. The tract for *pain sensations*.
2. The tract for *temperature sensations*, in which sensations of cold and those of heat must be separately regarded.
3. The tract for *pressure sense*.
4. The tract for *tactile sensations*.
5. The tract for *deep sensibility* (*bathyæsthesia*), among the components of which muscle sense with position, feeling, sensations of movement, etc., are most important.

Just as with motor spinal cord tracts, observers have endeavored to assign to each one of the known centripetal conducting tracts a function peculiar to itself alone. But here, too, it has been established with the increase of anatomic knowledge and the refinement of physiological investigation, more and more clearly, that several tracts may share the same function, and to a great extent, compensate for one another. Unfortunately, however, animal experiments relative to the conduction of sensibility, suffer from the difficulty in separating clearly the conscious sensations, rising in the cerebral cortex, from the general feelings evoked in the deeper centers of the brain.

The conduction of *pain sensation* in the dog and monkey is mostly given over to the lateral columns; their disjunction lowers it considerably, though it does not wholly destroy it. It is certain, at the same time, that the majority of the fibres concerned in the sense of pain, after their entry into the gray matter of the spinal cord, and especially of the posterior horn, crosses to the other half of the spinal cord, without, however, in the higher mammals a complete crossing of pain conductivity taking place. Disjunction of one lateral column, therefore, removes the pain sensation on neither side, but lowers it very markedly temporarily on the opposite half of the body. One can prove, furthermore, that the conduction of pain in the lateral column is, on the whole, confined to its ventral half. When the lateral columns are conserved, on the other hand, disjunction of the balance of the spinal cord columns leads to no disturbance of pain sensation. After destruction of the lateral columns in the dog, the preserved remainder of pain conduction, with which no exact localization is connected, and which probably represents only the subcortical components of the sensations of pain, just as it is retained

after the extirpation of the cerebrum, is in no way influenced by the disjunction of the posterior columns. On the other hand, the additional disjunction of the anterior columns in the upper cervical portion of the cord leads to an almost total loss of the pain sensation. But even now, if all the white spinal cord columns are severed at various levels of the cord, there still remains a remnant of pain sensation, induced by strongest stimulations.

If, therefore, the antero-lateral columns must be considered the most important tract in the conduction of pain sensations, it is, nevertheless true, that the anterior columns are also concerned, in this function, even if but slightly, or are at least able to assume this function by substitution. Finally, there must be considered the possibility, even if it be but a very slight one, of the sensations of pain being conducted through the short paths of the gray matter. This conception corresponds to the older discoveries in the rabbit, that the conduction of pain is not interrupted by a unilateral lesion of the spinal cord in a definite segment, and a second lesion of the other half of the spinal cord a few segments higher up. In the ape, on the other hand, such a bilateral hemisection of the spinal cord leads to complete loss of sensibility.

Since the discovery of *Gower's* ventral spino-cerebellar tract in the antero-lateral column, the chief conduction of the sensation of pain has been frequently ascribed to this tract. But since we know that *Gower's* tract ends in the cerebellum and that total extirpation of the cerebellum does not result in any disturbance of the pain sensation, this view must be rejected as irreconcilable with the facts. In the conducting path for the sensation of pain in the anterior column no continuous tract is concerned, but there is a chain of shorter and longer intersegmental nerve tracts, always returning to the gray substance of the spinal cord, the longest fibres of which, might, of course, commingle with those of *Gower's* tract (Fig. 56).

Concerning the *temperature sense*, its conduction in the lateral column corresponds in the main with that of the pain sensation. In the monkey there seems to be a bilateral conduction, showing a preference for the crossed side. But new experiments, carried out by *Kalischer's* training methods, have proved, that in the dog the conscious temperature sensations are throughout their course crossed. The training, connected with the temperature sense, was lost after severing of the crossed half of the spinal cord.

The conduction of the pain sense is paralleled by that for the *pressure sense;* when pressure is in any way increased, it is easily transformed into a pain sensation. Here, too, the main conducting path lies in the lateral column with preponderance of the crossed conduction in the spinal cord. In destruction of a lateral column in the dog, an increase of the pressure sense of the same side is often noted; it is accompanied also by a hyperalgesia. On the other hand, disjunction of the posterior column seems in no way to influence the pressure sense.

So far as the *tactile sensation* is concerned, it is naturally dependent upon a certain degree of pressure sense, and is lost, therefore, when the other is markedly lowered, as after bilateral disjunction of the lateral column. But, on the other hand, with retained or even increased pressure sense, the delicate localized tactile sensations may be severely lowered, or altogether lost. The

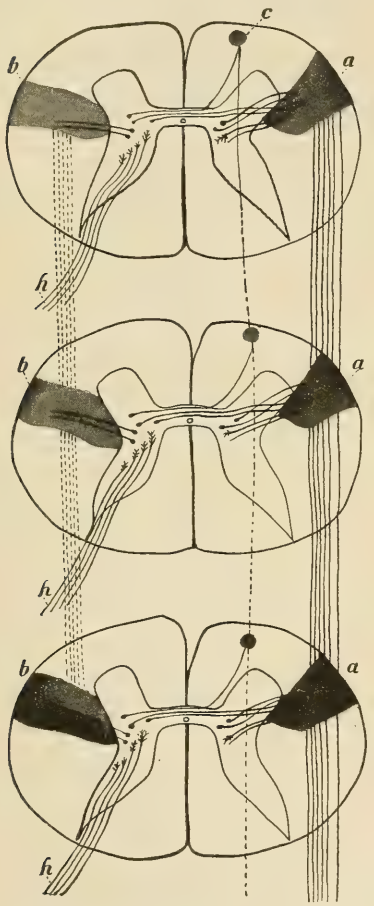


FIG. 56.—Conduction of the pain sensation in the spinal cord. *a*, Crossed antero-lateral column; *b*, antero-lateral column at the same side; *c*, crossed anterior column; *p*, posterior root.

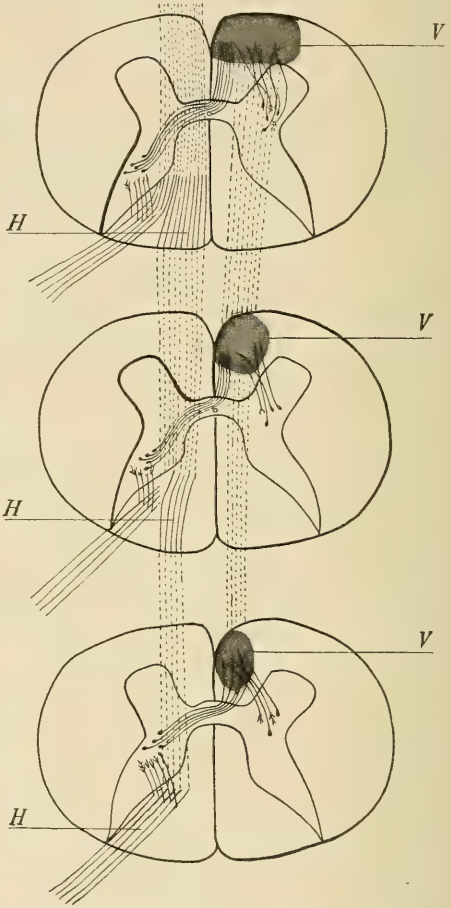


FIG. 57.—Conduction of the touch sensation in the spinal cord. *H*, Posterior column; *V*, anterior column.

conduction of the tactile sensations has been previously credited exclusively to the long posterior column tracts, which, without interruption ascend from the periphery and the spinal ganglia to the nuclei of the medulla oblongata and possess a secure connection with the cerebral cortex by way of the lemniscus tract and the optic thalamus. According to previous investigations, total division of the posterior column in the dog was said to destroy the tactile sensation, whereas, on the other hand, in abnormal heightening

of the sensibility, due to serious loss of blood, a rabbit, whose entire cord was divided with exception of the posterior column, still reacted to slight touch of the hind feet with movements of his head and ears. New experiments, on a dog, in which the total division of the posterior column, without additional injury, was established by exact microscopic investigation, have proved, however, that the tactile sensations are retained intact after total disjunction of the posterior cord. Nor does an extension of the unilateral division of the posterior column to the posterior horn and the extreme dorsal layer of the lateral column destroy tactile sensations in the extremities of the side concerned. If in the normal dog, one severs the anterior column in the first cervical segment, the tactile sensation is at first lowered, but soon returns to its normal degree. But if *total division of the posterior column is combined with that of the anterior column* in the upper cervical portion of the cord, the tactile sensations are completely lost, and the tactile reflexes disappear in spite of the intactness of the lateral column. At the same time the ordinary pressure sense and the pain sensations are in nowise disturbed.

There exists, therefore, a *double conducting path for the tactile sensations*, that through the posterior column on the same side, and that through the anterior column (probably only crossed). The anterior column tract of the tactile sensation must be built up in the spinal cord from shorter intersegmental tracts, since long anterior column fibres, ascending to the medulla oblongata from the lower spinal cord segments are not known. Both tracts are able to immediately replace each other, and both might, therefore, be normally used (Fig. 57).

For the *muscle sense* as well, the most important components of which are the sense of position and the co-ordination of movements, the posterior column was oftenest considered as the path of the conduction. But here too the isolated posterior column section in the dog results in no disturbance of the functions mentioned. In most recent times observers have given special attention to the spino-cerebellar lateral column tracts, since unquestionably a large number of sensations stream to the cerebellum from the domain of the deep sensibility, which are used up for the more delicate regulation of the equilibrium. If now these spino-cerebellar tracts, which occupy the margin of the lateral column, are completely divided in the dog, on one or both sides hypotonia results in the extremities on the same side and a disturbance in regulation of the principal movements in the roots of the extremities, which manifest themselves in standing, as well as in walking, as abnormal spreadings and overcrossings. These disturbances, which are suggested already in total division of the dorsal spino-cerebellar tract, appear in full severity only in total division of both spino-cerebellar tracts, the dorsal and the ventral. But all these pathological phenomena retrogress quickly, so that after four weeks, even with bilateral disjunction of these tracts, scarcely any disturbance is present.

Quite similar phenomena of loss of function of similarly transitory nature appear also after isolated anterior column disjunction in the upper cervical portion of the cord, so that we must ascribe also to the anterior column centripetal conduction for the deep sensibility, especially for the muscle sense. Especially severe is the appearance of these disturbances of the muscle sense in combined disjunction of the anterior and lateral columns; on the other hand, the intact lateral columns can almost completely take over all the innervations concerned here, after total disjunction of the anterior and posterior columns, followed by temporary disturbances.

It is probable, therefore, that in the conduction of the impulses brought together in the muscle sense, all columns of the spinal cord are concerned, even though under normal conditions the main conduction may be a function of the lateral columns. The larger number of impulses concerned here ascend in the same half of the spinal cord; but for the position sense, for instance, a bilateral path of conduction seems to be present.

If we again consider the *conducting paths of the centripetal impulses*, especially as they appear after the experiments on the dog, we must say, that the *posterior columns* have no specific, irreplaceable function. They are important factors in the conduction of the tactile sensations, and are probably able to take over a part, if only a small one, of the conduction for the muscle sense. The *anterior columns* are the second pathway for the conduction of tactile sensations, are feebly active in the conduction for the pain sensation the fibres probably ending subcortically, and take a not inconsiderable part in the conduction of the deep sensibility. The *lateral columns* finally are the main tracts for the pain and temperature sense, for the pressure sense and also for deep sensibility; only for the delicate tactile sensations, they possess no direct components, but can indirectly destroy them by very severe disturbance of the pressure sense.

Incidentally the curious fact must be mentioned here, that the posterior columns, the disjunction of which does not at all interfere with pain conduction, represent the most sensitive place (to pain) of the entire spinal cord—perhaps even of the entire central nervous system. Even the thoroughly anæsthetized animal starts up whimpering when the columns are pricked, and this is not because of tearing or cutting the root, which in fact is not nearly so painful, but there must be special endogenous pain tracts of the spinal cord, which course in the posterior column.

Complete division of an entire half of the spinal cord, in both dog and monkey is followed at once by *disturbances of the movements of the extremities on the same side*, which in the monkey are more serious and of longer duration than in the dog, but which in both animals disappear to a great extent in a few weeks, corresponding to the relations of the motor conducting tracts, as discussed above. So far as *disturbances of the various sensory qualities, after such a division of the spinal cord* are concerned, it is shown

by numerous competent experiments, that the muscle sense is disturbed much more on the side of the division and that the pain and temperature, as well as the pressure sense, are especially affected on the opposite side. But in the dog none of these forms of sensibility are destroyed by a half sided lesion of the cord, in fact, as a rule, they are generally restored almost to their normal condition in the course of time. Each of the two halves of the spinal cord, then, is able, to a certain degree, to conduct sensory impulses for both halves of the body. But even after two hemi-lesions of the spinal cord, which at a distance of several spinal segments, affect, now the right, now the left half, the sensory conductivity is not abolished, nor is it entirely destroyed by a third lesion a few segments higher, on the first side.

How do these results attained in the higher mammals, compare with those reached in *man?* Here we have, it is true, the immeasurable advantage, that by the help of speech much more exact information may be had about the sensations brought to consciousness; but, on the other hand, it is only seldom that definite focal injuries are observed with such clear phenomena of loss of function, as animal experiments offer us. Thus, for instance, in the posterior column affections of the tabetics, there is never mere loss of the posterior column fibres, but a combination with diseases of other posterior root fibres, which end in the gray substance, so that naturally no conclusions can be drawn concerning the functions of the posterior column from the phenomena of functional loss, observed in such cases. Also in tumors, which develop in definite parts of the spinal cord, for instance in the lateral column, one must be very careful in interpretation of the symptoms, because of their distal effects.

The best observations in *human pathology* for *establishing the conducting tracts of sensation in the spinal cord*, therefore, are presented by the *punctured wounds of the spinal cord.* These occur, as a rule, in young healthy individuals, and are comparable, in the simple division of definite portions of the spinal cord, with the conditions of the animal experiment. Here, however, there is an insufficiency of autopsy findings.

Punctured wounds, in which the knife enters the vertebral canal from behind, are followed, as a rule, by the appearance of the *Brown-Sequard symptom-complex.* Motility and muscle sense are lost on the side of the injury, pain and temperature sense, and, also to a greater or less extent, the pressure and tactile sense on the opposite side. On the injured side a slight tactile hyperæsthesia is noticed; also as a rule at the upper boundary of the anæsthesia, a small hyperæsthetic zone may be detected. In addition to punctured wounds with approximately clean division of half the spinal cord, this symptom complex, more or less pronounced, appears in cases of tumor, softening, hemorrhage, meningomyelitis of the spinal cord, which are localized preeminently in one-half of the spinal cord (Fig. 58).

We have already said above, that the loss of *motility*, appearing after

a unilateral lesion of the spinal cord, is capable of marked retrogression, especially in the lower extremity. So far as the *individual qualities of sensation* are concerned, the crossed conduction for the pain sense is undoubtedly far more developed in man than in animals. First, in all complete divisions of one-half the spinal cord and even of a lateral column, the pain sense of the opposite half of the body is lost, and limited restitution is

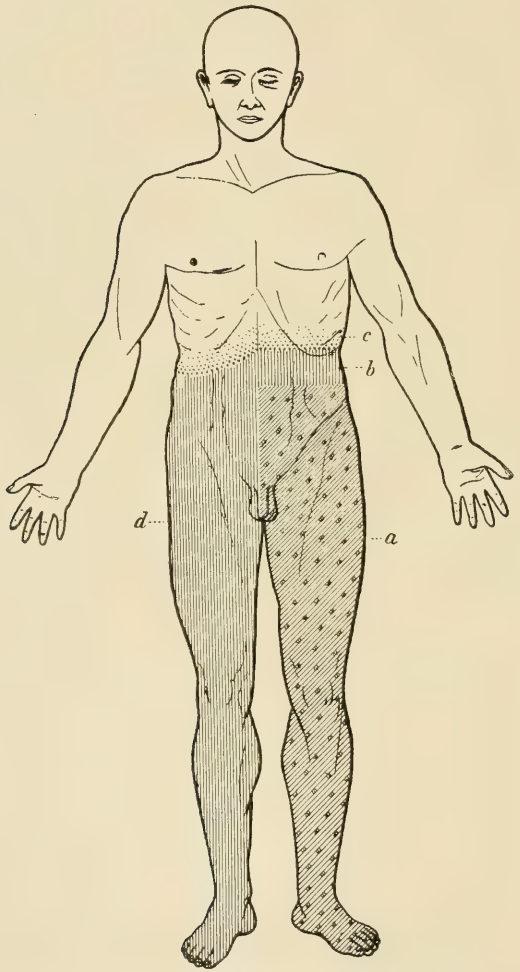


FIG. 58.—Left hemisection of cord. *a*, Motor and vasomotor paralysis, *b* and *d*, anæsthesia of the skin; *c*, hyperæsthesia of the skin. (*After Erb.*)

observed only after the lapse of considerable time. But there are a long line of observations on record, in which after a one-sided lesion of the spinal cord, complete loss of the pain sense was observed to continue for many years. But in all these cases the lesion seems to have extended to the other half of the spinal cord also. From the few cases that lived for years after a punctured wound of the cord, and whose spinal cord condition could then be

ascertained, it may be concluded with certainty that in man also, pain sensations, may be conducted though very incompletely along spinal cord tracts of the same side. Thus appears occasionally the symptom of *allocheiria*, in which, as the pain stimulus is carried only to the centers of the cerebral hemisphere of the same side and hence, as usual projected upon the opposite side of the body, the pain is not felt at the point of injury, but at the corresponding place in the other half of the body.

The *temperature sense* in man is conducted exclusively and normally, in the opposite half of the body, and as a rule does not seem to be restored, even after a unilateral lesion of the spinal cord has persisted for years. But we have at least one trustworthy observation, in which the preservation of the lateral column on the same side sufficed in spite of destruction of the entire opposite half of the spinal cord, to make an incomplete restitution of the heat sense possible after some years. Furthermore, several observations of almost isolated destruction of the antero-lateral column teach, that in man also, under normal conditions, the entire conduction for the pain and the temperature sense passes through this column. But all the objections that were quoted against the transference of this conduction to Gower's tract in animal experimentation, must be renewed here.

A separation of the *pressure sense* from the more delicate tactile sensations, has been accurately observed in only a few instances in man. At any rate, the pressure sense, far more than the pain and temperature senses, employs in addition to the opposite tracts, a less important system of conduction on the same side. For this conduction also, the antero-lateral column seems to take first rank. To what extent posterior and anterior columns may serve as compensating tracts, has not yet been definitely determined.

For *tactile sensations*, there is definite proof, from exact analysis of competent observations, that normally it must employ two tracts, one on the same, the other, on the opposite side. In many cases of unilateral lesions of the spinal cord, therefore, the tactile sensations are retained, while the pain and the temperature senses are lost. There is no doubt that one of the tracts for the tactile sensations courses in the posterior column of the same side. On the contrary, the frequently assumed hypothesis, that the second tract uses the opposite lateral column, is untenable in view of the observations presented; this second crossed tract must in accordance to the results of the animal experiments, pass through the anterior column. Therefore, in punctured wounds, in which the knife penetrates obliquely from behind, and in which, as a rule, one lateral and both posterior columns are destroyed, the tactile sensations are retained on both sides, since both anterior columns remained uninjured. That, on the other hand, the long posterior column fibres, that pass without interruption through the gray matter of the spinal cord to the medulla oblongata, conduct tactile sensation is proven by the retention of the latter in extensive cavity formations in the gray substance

of the spinal cord, though the pain and the temperature senses are lost (partial sensory paralysis).

If we turn, finally, to the *muscle sense*, we find that the *position sense*, about which alone we have exact knowledge, is at first lost in hemilesions of the cord, on the side of the lesion, but is, as a rule, considerably, even though not entirely, recovered. Here too, in man, the anterior columns are concerned in the condition as well as the spino-cerebellar lateral column tracts while the degree of participation of the posterior column is as yet doubtful.

The *Brown-Sequard symptom complex*, is, accordingly in man only in so far present as at first after a half sided lesion, the motility and the muscle sense are lost on the same side, the pain and temperature and mostly the pressure senses on the opposite side, whereas the tactile sensations from the very beginning have conduction upon both sides. But neither the conduction on the same side for motility, nor that on the opposite side for sensibility, is absolute. An extensive restitution of the functions through the other half of the spinal cord takes place, a phenomenon of the greatest importance for therapeutic efforts in human pathology.

If now this one sided lesion of the spinal cord affects the *upper cervical portion*, another series of phenomena of loss of function may be observed in addition. First *respiration* ceases on the side of the lesion, as the descending tract from the centers of the medulla oblongata to the phrenicus-center in the fourth cervical segment is interrupted. This cessation of the working of half of the diaphragm is permanent in man as well as in the dog and the ape. Yet, if, in the animal experiment, after a duration for weeks and months of the inactivity of one side of the diaphragm, the phrenic nerve of the other half of the diaphragm, that has continued its functions, be cut, and thereby this half also be made inactive, then, the paralyzed half immediately begins to work again (*Porter's* experiment). The respiratory stimuli, descending from the medulla oblongata in the sound half of the spinal cord, and which are now no longer able to reach the same half of the diaphragm, make a commissural path, at the level of the fourth cervical segment, to the phrenicus center on the other side, thus awaking it to renewed activity; a peculiarly instructive example of possible processes of restitution in the central nervous system. These spinal respiratory paths pass, as has been proven, by attempts to sever them in the dog, for the most part in the ventral part of the antero-lateral column, to a lesser degree in the lateral part of the anterior column.

Furthermore, one observes in both animal experiments and in man, after a high half sided cervical lesion, an increase in cutaneous temperature on the side of the injury, which is said to be due to a paralysis of *vasomotor tracts* as they pass from the medulla oblongata to the spinal cord, coursing more than likely, also, in the antero-lateral column.

Finally, let us recall once more the *oculo-pupillary symptoms* after a half sided lesion of the cervical portion of the cord. They consist of narrowing of the palpebral fissure, of contraction of the pupil and occasionally also, of a retraction of the eyeball (enophthalmos), and are due to a severing of the sympathetic fibres, passing from the medulla oblongata to the oculo-pupillary center in the first dorsal segment. Here, too, a definite restitution of the nervous conduction is apparently possible, through the other half of the spinal cord.

Concerning the many finer details of diagnostic importance, especially with regard to the lowest segment of the spinal cord, we must refer the reader to the separate chapters on the pathology of the spinal cord.

2. SYSTEMIC DISEASES OF THE SPINAL CORD

(a) Tabes Dorsalis

(Ataxie locomotrice progressive.—Locomotor ataxia.—Rückenmarkschwindsucht)

BY

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We designate as tabes dorsalis, or *tabes*, the most frequent of the systemic degenerations of the spinal cord, the so-called gray degeneration of the posterior columns and the posterior roots. This degeneration in general, must be considered as progressive and incurable. It offers a great mass of classic clinical symptoms, among which those of the motor sphere, with the exception of ataxia, occupy a very insignificant position. On the other hand, the subjective and objective disturbances of sensation—and, indeed, of almost every quality thereof—those of the reflexes, of the sensory and trophic functions, play the main rôle.

The *ætiology* of tabes is—at least in the way the question is usually formulated—to be regarded as sufficiently clear to us to-day. We do not know the "excitant factor of tabes," if there is such a one, but we do know that the disease almost always appears only in people who have had syphilis at one time. In the great majority of cases it is a meta-syphilitic disease. This states the main fact about its ætiology; we must, however, enter in somewhat more detail upon the theme, that has been the subject of ardent dispute for decades.

The ideal proof of the connection between syphilis and tabes would be established, if we could only succeed in proving, the same excitant clinically in both diseases. We may consider, to-day, as the cause of syphilis, the *Schaudinn-Hoffmann* spirochæta pallida; but it has not yet been possible to find in the cerebro-spinal fluid withdrawn by lumbar puncture or in the central nervous system of (pure) tabetics, spirochætes (this, of course, because of the nature of tabes as a metasyphilitic disease, was a priori to be

supposed improbable). [Very recently the spirochætes have rarely been found.] The proof of the presence of syphilitic anti-bodies in tabetics (*Wassermann-Plaut* reaction) has indeed recently been successfully shown with comparative constancy (*Citron*), but, in the first place, there is as yet but little clinical material and, secondly, we must lay emphasis on the fact that the reaction (which strikingly appeared rather infrequently in the cerebro-spinal fluid, and mostly in the serum) proved that the tabetic very probably (even when he denies it) has at some time had syphilis, but does not at all prove that tabes depends upon syphilis, since most non-tabetics, who had been infected with syphilis, similarly showed anti-bodies. Besides these direct methods, pathological anatomy also fails, as it, as we shall see below, shows the lack of really *specific* changes (in so far as the word "specific" may be used here) in tabes.

For the present, therefore, the statistical method of the proof of the existence of syphilis remains the most serviceable for our purposes. And this teaches clearly and indubitably that

1. Almost all tabetics have at one time had syphilis, whereby of course an exact anamnesis (or circumstantial evidence in certain cases) must often suffice as proof. The largest existing table of statistics, collected by *Erb* (1904: 1100 cases) gives about 90% of syphilitically infected tabetics; the French writers (*Fournier, Déjérine, Belugon and Faure*) 77 to 97% of former syphilitics, *Strümpell* 90%, *Gowers* 75 to 80%, *Negro* 89%, *Homén* 83%, etc. The number of authors crediting only a small percentage to syphilis (*Storbeck and Gutmann*), grows constantly smaller and smaller, and we may accept the fact to-day as established, that *approximately* 90% *of all tabetics were formerly infected with syphilis.* All "proofs to the contrary" constructed partly at least artificially, serve in objective judgment only to confirm these results. Of 10,000 non-tabetic patients, only 21.5% had syphilis in their anamnesis (Erb). The fact that of all syphilitics only comparatively few get tabes, proves that tabes is a *rare*, and not a frequent sequela of syphilis.

For the anamnestic proof of syphilis, it is of course essential, that the physician knows "how to ask questions." The unpleasant fact of a past experience with syphilis is an admission that nobody cares to make; very many patients (especially of the lower classes, women, etc.), are not even acquainted with the fact of having had syphilis or have forgotten it, either because it was but slightly treated or not at all (this appears with extraordinary frequency in the case of those who become tabetics later or because its course was brief). Then one must seek for indications: eruptions, swelling of the glands, infections of members of the family, abortions in the wife or sterility, relapsing cerebral paralyses, etc., in the history, leukoderma, plaques in the mouth, scars of all sorts, in rare cases also for typical tertiary diseases, gummata, etc., in the physical examination. In tabes of children

(rare) one must be on the watch for extragenital infection or for congenital syphilis. The search succeeds more easily with men than with women, and more easily in the upper than in the lower classes. Certain occupations, which seem peculiarly to increase this liability to the acquisition of syphilis, exhibit the most tabetics (merchants, travellers, actors, officers), others, correspondingly the fewest (clergymen). The history of the individual peculiarities of the infection (*only* hard chancres, eruptions, condylomata, etc.), varies considerably; in general, it seems, as has been remarked above, that the majority of apparently slight and but little treated cases lead to future tabes. The *time* of the appearance of tabes after syphilis is also somewhat variable, but it appears scarcely ever earlier than 5 or later than 20 years after the infection; in late infection, also, the outbreak of the spinal cord disease can be established, occasionally, only close to old age. Tabetics are, moreover, nearly always immune to syphilis. Tabetic couples, i. e., when husband and wife, one after the other, are attacked by tabes, are syphilitic, almost without exception (*Fischler*). Why in certain races which are at least in part fairly saturated with syphilis, tabes appears but comparatively seldom, has not yet been completely explained (cf. below). (Tabes was never seen in the American Negro until recent years. To-day it, and paresis, are becoming common. *Burr.*)

2. The remaining ætiological possibilities worthy of any consideration—compared with syphilis—are of but slight moment. In this matter, however, the statistics are unfortunately but slightly satisfying, since only a few writers have objectively considered all such possibilities with complete impartiality. *Erb* has done this most thoroughly (cf. below). As to these remaining factors, it appears that the *poisons and infections* that are worth mentioning, outside of syphilis (mercury, alcohol, tobacco, influenza, gonorrhœa) with the exception of gonorrhœa alone play an insignificant rôle; according to Erb, 90% of the tabetics had gonorrhœa; but for the most part they were those, who at some time also had suffered from syphilis. *Sexual excesses,* in so far as can be ascertained, also play a small part; on the other hand, *traumatism* may give rise to tabes under certain conditions. "*Catching cold,*" on the contrary, the "rheumatism" in the anamnesia, is, in a disease, that begins so frequently with pulling and tearing pains, to be regarded skeptically and actually can be but seldom demonstrated on closer investigation. The *neuropathic disposition,* the "ectodermal weakness of the germinal layer" (*Bittorf*) doubtless co-operates, though we must note, that genuine signs of physical degeneration in the patients are, in our experience, of less frequent and regular occurrence, than nervousness in the family. Finally, we must mention the theory of tabes as the "exhaustion disease" (Aufbrauchkrankheit) of the nervous system (*Edinger*). In physical overexertion, particularly where the activity of the individual is limited to one side of the body, there takes place according to *Edinger* an early using up of the nerve substance

which may develop into systemic degeneration of the tracts concerned. This holds good, incidentally, for nervous regions, weakened by other kinds of diseases, poisons, etc., and thus especially for the spinal cord of tabetics, which has been influenced by syphilis. The hypothesis is quite illuminating, and holds good for many tabetic symptoms (thus, for instance, after extraordinary exertion of the legs, as in the case of Marathon runners, it happens not infrequently, that the patellar reflexes do disappear, but they *remain* absent for a long time or even permanently, only in those individuals who have had syphilis), but the theory does not explain, why, among the countless individuals who have had syphilis, and who have frequently undergone one-sided physical over-exertion, tabes does not appear far more frequently than actually happens.

Only *Erb*, as we have said, has made exact investigations of the conditions having a possible ætiological relation to tabes, which are worthy of consideration and has put them together in tabulated form. It appears, naturally, that very frequently several ætiological factors co-operate; but if one counts only the cases, in which *only one* ætiological factor was proved to be possible (with the high percentage of syphilis in anamnesia, this number is naturally small) one finds in cases of

Syphilis alone	27.0%
Neuropathic heredity alone	0.7%
Cold alone	1.4%
Over-exertion alone	0.3%
Sexual excesses alone	1.0%
Traumatism alone	0.3%

As to the *manner of the working* of all these evil influences, we have unfortunately, up to now, only conjectures; the majority, it is probable, furnish only the *exciting* factor for the development of the disease. But how the spinal cord becomes, primarily, less resistant or more receptive to the exciting stimulus, we do not know. This question is most important as to syphilis: Is it only the most frequent "agent provocateur," or the real cause of tabes? Many observations speak for the latter view. I must mention also the hypothesis of French authors of a special "nerve syphilis," a "syphilis à virus nerveux," which causes but slight phenomena to appear on the skin, mucous membranes, etc., outside of the primary chancre, but lays the stress of its dire work to the destruction of the nerve elements, upon which is based the future tabes or paralysis. Many striking phenomena, as the occurrence of tabes in married couples, the frequency of cases of tabes among syphilitics whose infection came from the *same* source (*Brosius*) might thus be explained.

Another, at first sight amazing hypothesis (*Loewenthal*), assumes that not syphilis itself or its toxins, but the protective substances (anti-bodies)

produced years after the infection, cause tabes. This, in any case, extremely doubtful hypothesis is to be merely mentioned here; it proves only, as does all that has been said above, that though we know that *syphilis is the main factor in the ætiology of tabes*, we do *not* know *the manner of its working*.

If we turn to the *pathological anatomy* of tabes, things are simple enough so far as regards *histology*, more complicated as to the *distribution* of the process in the central nervous system, and extraordinarily complicated and difficult to explain so far as *pathogenesis* is concerned.

If we summarize our present knowledge on these points, we may say: *Tabes is a degenerative disease, characterized by progressive destruction of the nerve fibres and loss of the medullated sheaths as well as by a secondary increase of the glia*—so-called gray degeneration. *It is located in the posterior roots and the posterior columns—first in the column of Burdach, then in Goll's column—and in a few less constant points. It begins probably with a specific syphilitic inflammation (in the broadest sense) in the posterior roots and a chronic slight meningitis, which—segment after segment being affected—gradually causes the tracts of the central organ concerned to degenerate.* We shall try to prove these statements in the following.

The **histological** process may almost be described as banal, so typically does it repeat itself in the majority of cases. "The nerve fibres degenerate, lose their medullated sheaths in a more or less irregular way, are in part thickened and swollen, later atrophy, together with the axis cylinders, and finally disappear altogether. The proliferated glia appears as a delicate fibrous network, in which, frequently, the gaps left by the destruction of the nerve fibres are still visible; later it becomes transformed into a delicately waved, striped, tough mesh, in which nuclei and neuroglia cells, as well as corpora amylacea are present in great numbers. The vessels are but slightly changed; have thickened and hyaline walls, narrowed lumen and here and there a somewhat richer intercalation of nuclei. In quickly progressing recent cases granular cells have also been found (*Erb*)."

Changes quite analogous to these are found in the posterior roots, and especially in the part that lies between the entrance into the dura and the spinal ganglion, which French writers call the "nerf radiculaire," since *Nageotte* wrote about it, and here and there, but *not constantly* in the spinal ganglia themselves. This division of the roots shows interstitial neuritis, and later, degeneration; according to the most recent investigations, there appear, in the beginning, regenerative processes also (*Nageotte, Marinesco*), especially in the form of "collateral regeneration," but the process always ends in total destruction of the fibres. Finally, the meninges are frequently also—constantly according to French writers—affected by diffuse proliferation of the cells, especially in the walls of the veins and the arteries of the meninges, which, according to *Nageotte* and others, is identical with the

changes in the vicinity of the vessels, observed in genuine syphilitic processes (these cells are also found in large quantities in the cerebro-spinal fluid of tabetics). The *slight* meningitis—in contradistinction to which there exists a rarer coarse, chronic meningitis, with thickening and adhesions—occurs in the vicinity of the posterior columns and the points of entry of the posterior roots. It is seldom that the anterior roots show a similar degeneration; on the other hand, we find frequently, in addition to the changes in the fibres, degeneration in the ganglion cells, and especially in those of *Clarke's* columns, rarely, also in those of the anterior horns (the latter only in isolated forms of tabes with amyotrophy, arranged in nests or foci, *Lapinsky*). Occasionally, also, the degeneration affects peripheral motor and sensory nerves, but almost only in the cases, in which, clinically also, the simultaneous presence of neuritis, on account of existing pareses, etc., may be recognized.

It must be emphasized, however, that the changes in the "nerf radiculaire" and the meninges, in the form described, are an outcome of French investigations of recent years, and are still awaiting full confirmation; but they have been found with such constancy by these investigators, that, considering their importance for pathogenesis, we are justified in regarding them as peculiarly significant.

Macroscopically a tabetic spinal cord can often be easily recognized by the small volume, the "disappearance" of the posterior columns; furthermore the portion of the posterior columns affected is characterized by the easily recognized gray discoloration, not only when the cord is intact but more particularly on cross-section.

These are the real tabetic changes. Besides these, we find, not very rarely pathological pictures which belong to other diseases of the central nervous system: combined diseases of the columns (with participation of the lateral cerebellar tracts, the pyramidal tracts, etc., cf. below), proliferations in the molecular layer of the cerebellum, formation of cavities (combination with syringomyelia), and genuine gummatous, syphilitic processes of the meninges, of the substance of the brain and spinal cord; finally the frequent combination with progressive paralysis, even from an anatomic standpoint.

How, then, does *the tabetic process spread?*

In part the localization has been already mentioned: the membranes, posterior roots, posterior columns. In detail, we can start from the degeneration of the posterior roots, even when they do *not* show the earliest and most marked appearance of the process, as occasionally happens. But if we assume that the process starts regularly from the posterior roots (cf. below), we should have to see the degeneration of the fibres of the posterior roots pass over into the posterior columns, and, according to the level of the spinal cord, affecting more either the columns of *Burdach* or those of *Goll;* it would have to rise, in part, to the nuclei of the posterior columns in the

medulla oblongata, and, for the rest, pass over into *Clarke's* columns of the posterior horn and the *Lissauer* marginal zone. Also the descending fibres in the posterior column would have to undergo degeneration. Then, surely, the process could often be traced up to the spinal ganglion on one side, up to the cortex of the cerebellum, into the inner capsule and to the sensory centers of the cerebral cortex.

This actually corresponds to the course of the disease in the majority of typical cases, with the one restriction mentioned, that the first *clearly visible* changes, appear, as a rule, not in the posterior roots, but in the posterior columns. In fact the first change observable in the lumbar segment is a symmetric field of degeneration in the columns of *Burdach*, laterally next to the posterior horn, the very place of the "bandelettes externes," the entry zone of the roots. This is the typical condition at the initial stage of the disease. Thence the process continues upwards. Corresponding to the course of the columns of *Burdach*, with the degeneration of every higher segment, the disease moves *more towards the midline*, finally into the columns of *Goll*. The higher we go in the spinal cord, the more clearly are the columns of *Goll* affected (in regular sequential succumbing of the individual segments), so that we can lay down this rule: in the lumbar segment, first the columns of *Burdach*, then the total cross-section of the posterior columns are attacked; in the dorsal and cervical segments, first the columns of *Goll*, only later the entire cross-section; in the medulla, the nuclei of the posterior columns in the vicinity of the calamus scriptorius. As a rule, the *Lissauer* marginal zone and *Clarke's* columns of the posterior horns, are affected at the same time, the latter, occasionally, in such a way that the cells remain intact, whereas the fibres degenerate. The posterior roots themselves are also always affected, even if not always apparently to the same extent, as the posterior columns, so that many authors assume an "elective disturbance" of the root fibres. Very often we find cell degeneration in the spinal ganglia, even if, and this must be emphasized, not absolutely constant. On the other hand, in the lumbar segment, the dorso-medial bundle, and farther up, the so-called ventral posterior column field, are almost invariably spared.

Concerning the participation of the meninges, which may be described as rather constant, we have spoken above under histology. Frequently we find also affected the reflex collaterals, radiating into the anterior horn, in the rare cases of combination with spinal muscular atrophy, also the ganglion cells in the anterior horn. Not rarely, the gray degeneration attacks also *Gower's* bundle and the lateral cerebellar tracts, but seldom, on the other hand, does it affect the pyramidal tracts. In the cortex of the cerebrum and of the cerebellum we occasionally find similarly degenerated fibres. The peripheral sensory fibres (rarely the motor ones) show, at times, a degenerative process, which can not, however, compare in severity with

the columnar degeneration. Very much more frequently, however, are the cerebral nerves attacked, and especially, and almost regularly, the *opticus*, in the sense of a primary parenchymatous degeneration, possibly omitting a small bundle (*Marie* and *Lèri*), and this degeneration seems to be quantitatively independent of the simultaneously occurring disappearance of ganglion cells in the retina. Next in frequency the abducens, the oculomotor and trochlear nerves are affected, and, far more rarely, the auditory, olfactory, trigeminal. Positive findings in the sympathetic have been strikingly rare (cf. below).

In contradistinction to these more or less regular systemic degenerations in the normal course of tabes stand some apparently irregular cases, in which, for instance, there are scarcely any changes in the lumbar segments, but serious ones in the posterior columns of the cervical region (for instance, in so-called tabes superior, only in the columns of *Burdach* of the cervical segments), or where spots of degeneration are scattered arbitrarily, as it seems, over the posterior columns. If we follow up these cases, we find almost invariably that the cause is an irregular sporadic lesion of *individual* posterior roots, *individual* segments, which can even, apparently, be confined to *one* segment only. From all this it appears with considerable definiteness, that in tabes we have a *segmentary*, *progressive*, normally *ascending disease of the posterior root regions*, that is, *a lesion of the posterior columns of a radicular type* (*Erb*). Upon this pathologic-anatomic fact, *Nageotte* and other French authors base what is, at present, the most plausible *theory of the pathogenesis* of tabes, into which we shall enter briefly.

According to *Nageotte*, the process is as follows: At first, there develops a diffuse leptomeningitis, which, because of the small-celled infiltration of the pia and the changes in the vessels, which are supposed to strongly resemble syphilitic changes, he regards as specific-syphilitic. Serious local changes, adhesions, etc., do not arise from this (cf. above); on the other hand, there soon occurs a root neuritis (cf. above, a neuritis of the part between the dura and the spinal ganglion). Hence the degeneration occasionally extends to the spinal ganglion, but always the entry zone of the roots and the posterior columns, and is even, at times, most clearly visible in the posterior columns, since, according to *Nageotte*, the collaterals or the so-called short root fibres may degenerate first at the distal end. That from the meninges, the nerf radiculaire is affected with special ease, is explained by the indubitable fact, that lymphatic channels pass constantly along the posterior roots, which can bring the supply of syphilitic and other poisons to bear with peculiar intensity upon the restricted space.

This theory, doubtlessly, meets most of the demands. It has only two weak points. One, that the *syphilitic* nature of the meningitis has thus far not been established, since we simply do not know any changes, which must be regarded as certainly syphilitic and which can be proven such experiment-

ally; second, that up to the present time, there have been too few special anatomic investigations of the nerf radiculaire.

The assumption of a primary disease of the *spinal ganglia* (*Marie* and others) is weakened by the fact that the changes demonstrated are too inconstant; nor is the exceedingly different participation of the incoming and outgoing nerves favorable to the theory.

Likewise the assumption of a diffuse toxic influence acting on the posterior columns through the poison circulating in their lymphatic channels, coming from a meningitis "posterior" luetica (*Marie* and *Guillain*) is a mere hypothesis.

Leyden and *Goldscheider* believe the process to have its beginning in the peripheral sensory nerves, but they are too rarely found affected to make this supposition probable.

The possibility that a primary toxic disease of the intramedullary root tracts exists at the outset, which is the theory of *Strümpell* and others, is, on the other hand, not to be set aside lightly.

To what extent the results of the *investigation of the cerebro-spinal fluid* have been favorable to the one or the other hypothesis, we shall see below.

Now the question arises; *are these pathologic-anatomic processes*, which have been described, *of syphilitic origin or not?* It is impossible for us, at this time, to answer either in the affirmative or the negative. The reason for this is, that the means for certain diagnosis of a specific syphilitic change, is as yet denied us, so that it has been possible, in recent times, to doubt even the specific character of the genuine gummatous changes. So long as we can not isolate the excitant of syphilis, or its toxins from the regions affected, or experimentally evoke a typical tabes, by the use of syphilitic matter, we shall have no positive proof. To-day we can only say, that, according to the status of our present knowledge, it is possible, but that it has thus far not been proven, that the changes of the nervous system found in tabes are with regard to the pathological anatomy of a specific syphilitic character. But the observations, attesting the correctness of this supposition, are multiplying. The discussion of the specific tabetic anatomic changes in the rest of the body (arthropathies, etc.), will follow their mention under symptomatology.

The **clinical symptoms** of tabes are unusually varied. Their distribution into stages of the disease (initial or preataxic stage, ataxic stage, paralytic or final stage) is correct only in isolated cases, because of the differences in the course of the disease, and is subject to the point of view of the observer. Most easy to demarcate is the *initial stage*, the duration of which, however, may vary from a few months, or even weeks to 22 years (mentioned by *Erb*). But it is characterized by symptoms which belong regularly to this stage.

Since it is usually only by chance that the diagnosis of tabes is made in this early stage, the possibility of its demarcation depends often upon the statements of the patient, who has already reached the later "recognizable"

stages of the disease. The most constant complaint of these patients to the physician is, "I have had rheumatic pains for so many years" (also often, alas, in the form "my doctor has been treating me for years for "rheumatism"). This "rheumatism" is with tabetics the almost invariable, deceptive designation of the *lancinating pains*, which as a matter of fact are most commonly the first symptom of the disease. Almost all tabetics give the same description of this disturbance: *lightning-like shooting* pains, lasting from an instant to several seconds—very seldom of longer duration—which are unusually violent, so that the patients often scream aloud. As a rule, these pains do not appear regularly distributed over long periods of time, but rather massed, after exertions, excesses, psychic irritations, exposure to cold, change in the weather, occasionally, too, without any apparent cause. They usually follow the single nerve trunks and the localization of the disease in the spinal cord and are usually, therefore, observed first in the legs (sciatic, crural), only later in the arms and the trunk, very seldom in the head. By no means, however, do the pains always recur in the same nerve, but occur within the same exacerbation period in all possible regions attacked; but the contrary also takes place at times, and in these latter cases there is often hyperæsthesia to touch, pressure, etc., in the regions concerned. The lancinating pains are present in 90% of all tabetics (*Erb*) and not infrequently characterize the entire picture. There are patients, who, through these terrible pains *alone*—having very few other symptoms—are completely deprived of their ability to work and their joy in life and finally commit suicide. Very many more, even when these pains finally disappear after many years, have in the meantime become morphine victims. It is remarkable that besides the crises (cf. below) and these peculiar lightning-like pains, other pain is not frequent in tabes.

Paræsthesias, the subjective sensory disturbances, are considered as the next early symptom. Many tabetics complain of their feet "falling asleep," "feeling prickly," of a sensation of "ants crawling over their feet," and, moreover, this disturbance (not taking into consideration the rare cases of tabes superior, cf. above) is frequently a typically ascending one. It begins in the tips of the toes, separate toes become "dead," then the soles become paræsthetic, or the one margin of the foot, or the entire foot. The feet are continually cold or warm. Then the paræsthesias become stocking-like, the feeling of a band about the knee arises, which develops later and higher, as the "*girdle feeling*" in the trunk, to one of the classic tabetic paræsthesias, which often appears as an early symptom. The ascending type, which appears also in myelitis, is occasionally lacking; the patients complain of their forearms going to sleep (ulnaris sensation), of the same feeling in the rectum, scrotum, etc. A feeling of heaviness in the legs is to be interpreted partly as a pure paræsthesia, partly as a genuine *diminution of muscular strength*. This also is very frequent in the initial stage as *subjective* dis-

turbance. The patients do not like to take long walks; they *tire quickly*. At this time also there usually appear disturbances in the sphincters and of the sexual function. In the beginning, *disturbances of the bowels*, because of the great frequency of bowel sluggishness, are rarely of much value (*incontinentia* alvi can scarcely occur as an early symptom); on the other hand, one often hears that the patients have to urinate more frequently, that they can not properly hold their urine, or (more seldom) that they have to exert strong pressure in urination (incontinentia vel retentio urinæ). Male patients complain almost invariably (80%), and usually even in the early stage, of *impotence* in the widest sense; as a rule, at first, of insufficient erections, premature or insufficient ejaculation; the loss of the libido, to the torture of the patient, usually follows only later. In women, similar changes seem to take place. Frequently the stage of impotence is preceded by one of increased libido, occasionally, too, of increased sexual capacity.

As the last preataxic symptom of a subjective nature, we note disturbances affecting the *eyes;* most frequent is diplopia which often appears first as "seeing dimly," as a confused picture, etc. Occasionally it persists as a constant symptom, but it usually disappears, yielding to other phenomena; it frequently, however, returns. Other patients complain first of a decrease in the vision, which affects usually one eye at the start, later becomes bilateral, and may lead to complete amaurosis. There are tabetics, who have absolutely no symptoms, as long as they live, except total blindness and possibly disturbances in the tendon reflexes.

For these ocular phenomena, we find even in the early stage, *objective* foundations; disturbances in the muscles of the eyes and in the opticus, as well as pupillary rigidity are present. Corresponding to the diplopia just mentioned pareses of the abducens (most frequent), oculomotor (ptosis also very frequent) and trochlear are often temporary in the beginning, their intensity changing in the course of weeks, even of days; they may disappear completely and never return. But often in the later stages there develop permanent, usually total, paralyses; we find ophthalmoplegia interna (paralysis of all the *inner* muscles of the eye), externa and total ophthalmoplegia, in which, back of the sunken lid, the ball usually slightly turned inwards and downwards is fixed immovably (Fig. 59).

Changes in the optic nerve are frequent in the ophthalmoscopic picture, but the findings are fortunately not always parallel to the actual visual disturbance. The disc is more or less grayish-white, in later stages often discolored to a porcelain white, thus marking the presence of *atrophy of the optic nerve;* the vessels of the disc are contracted, its margin marked with unusual sharpness; the retina is, as a rule, unchanged. In cases, in which the discoloration from gray to white proceeds very rapidly, there appears also a progressive loss of vision, finally leading to blindness; on the other hand, however, one sometimes finds in tabetics, who complain only of slight visual

disturbances, pure white discs in the final stage. Characteristic is the increasing contraction of the visual field, especially for colors, which may lead to a partial or total color blindness long before the appearance of total amaurosis.

By far the most frequent of all changes in the eyes, however, is *pupillary rigidity*, the *Argyll-Robertson phenomenon;* it appears in almost all instances even in the early stage and is associated with sluggishness of reflexes. Each eye is tested separately by allowing daylight or concentrated light to fall on the previously covered eye; the pupil contracts slowly, sometimes irregularly, often not at all. Next we allow the patient to converge, or to

FIG. 59.—Total paralysis of the oculomotor on both sides. Attempt to raise the lids (to knit the brows) prevented by ptosis.

accommodate for distance; in both cases a distinct contraction is visible. Therefore, there is pupillary rigidity to the stimulus of light, whereas the convergence and accommodation reactions are retained. This is the rule, occasionally even the only early symptom in tabes. As a rule the pupils are usually of unequal width in the beginning (anisocoria), one usually partly dilated or mydriatic, the other in many cases and at a very early date considerably narrowed (to the size of a pinhead); not seldom they are no longer round, but elliptic or angular (without the presence of posterior synechia). Also the dilatation reflex of the pupil by means of sensory stimuli (pricking the skin of the cheek), is absent usually in advanced cases.

We mentioned above the subjective *sensory disturbances* of the early stage. One of them is, as a rule, even now the expression of an *objective* disturbance: the insufficient feeling of the patients relative to what is beneath their feet. They do not know whether they walk on carpets or boards, on clay or stone; they have the feeling of "walking on velvet," etc. Then we find mostly a dulling of tactile sensations, probably also of the pressure sense, whether it be in the plantar surface, in the outer or inner margin of the foot, or in the toes. Also hypalgesia and analgesia occur, and, likewise, we find early a slight degree of *hyperæsthesia to cold* in the trunk, which may be subjectively represented by the statement, that washing with cold water, to which the patient has been formerly accustomed, has become unpleasant or even painful.

As the last of the actual early symptoms, I mention the changes in the *tendon reflexes*. Only very seldom do we miss in the initial stage a weakening or *absence* of one or more tendon reflexes. Of course, to establish this, it is invariably necessary to carefully test all the more important tendon reflexes of the body, above all—after the patellar reflex—the Achilles tendon and the triceps reflexes. Absolute relaxation of the muscles and joints concerned in the test is the necessary condition, before one can speak of a reflex being absent (for the methods of testing the reflexes cf. under "general diagnostics"); at every such examination the *Jendrassik* or other similarly valuable method for the purpose of facilitating and increasing the relaxation must be employed. If these precautionary measures are employed the earliest symptom in tabes is found to be a *difference* between two symmetric tendon reflexes, the one being usually normal (scarcely ever exaggerated), the other diminished. And, moreover, this diminution, which quickly increases to a "total absence" of the reflex, by no means always affects the patellar reflexes first (*Westphal's* phenomenon), but very often the Achilles tendon reflex, or in tabes superior the triceps reflex.

If we mention, furthermore, the occasional appearance in the beginning stage of the Romberg phenomenon, to be discussed at greater length below, we shall have mentioned in a nutshell the usual *early symptoms of tabes*, to which other authors (among them, Erb) it is true append a long list of others. Meanwhile, the borderline is always more or less arbitrary, unless importance is attached to the appearance of *ataxia*, which is often absent at this early stage. The initial stage often passes unnoticed into the fully developed disease, which we must now discuss, connecting the ataxic with the so-called final stage.

As mentioned above the *pains* may accompany the unfortunate patient through the entire course of the disease. They may become less frequent at the height of the fully developed tabes, but they scarcely ever disappear entirely and frequently, as "tabes dolorosa" form, at least subjectively, the most prominent symptom. But in additon to the lancinating pains, the radius

of action of which naturally increases with the progress of the disease, and which to-day attack one part of the body of the patient, tomorrow, another, we meet also with an entirely different kind of pains—the *crises*.

The term crisis includes painful sensations of a certain "degree," mostly also of a somewhat longer duration, which almost invariably proceed from organs, or are at least localized in them, the innervation of which is derived also from the sympathetic in addition to the cerebral and spinal nerves (especially of the vagus group). This naturally does not mean that these pains are transmitted through the sympathetic—we shall speak below at greater length upon this point. But it is nevertheless striking, that the organs most frequently affected (stomach, bowels, kidneys, etc.) according to the prevailing views (*Lenander, L. R. Müller, and others*) and investigations, are susceptible in but a slight degree to sensory stimulations. The first place is beyond a doubt to be assigned to the stomach, or *gastric* crises.

The typical gastric crisis begins almost without any warning, rarely with slight disturbances of the stomach, loss of appetite, etc. This is followed by unusually violent pains in the epigastrium, which resemble the pains of gastric ulcer, but may become more colic-like; they are mostly described as tearing, jumping, boring. Almost simultaneously, the patient suffers with uncontrollable vomiting, first of food, then of mucus, which is mostly stained with bile, and usually shows but slight acidity. The patients vomit then throughout the duration of the crisis—usually for a few hours, but also with interruptions for days and weeks—literally all the nourishment they take, so that occasionally artificial feeding must be resorted to. In the crises pain and vomiting coincide, but the attack of pain may persist in the intervals between vomiting. The loss of strength of the patient is enormous during this time, but as recovery from the crises, which always end suddenly, is just as quick, the conditions are rarely so bad as to menace life. The stomach crises are the predominate features in many cases of tabes and recur every few weeks or months, but, as a rule, only during a period of the disease, which may however last through several years. The majority of tabetics are fortunately spared them, but the percentage of those affected is not inconsiderable.

The other forms of crises occur far less frequently.

In "*intestinal crises*," there appear at intervals profuse attacks of diarrhœa, usually with colic-like pains.

Attacks simulating renal colic have been described as ***kidney-crises***. *Crises of the bladder, urethra, testicles, clitoris,* are all analogous conditions, which the patients localize in the organs mentioned. As a rule they are of much shorter duration than the gastric crises; those affecting the genital region are frequently accompanied with intense feelings of libido, as well as with pain.

Somewhat more frequent are the *laryngeal crises*. The onset is characterzed by an apoplectiform coughing fit, with spasm of the glottis, which may

lead to serious attacks of choking, and often to temporary loss of consciousness. This form is of all those mentioned, the only one that is directly dangerous to life. Attacks of *tachycardia* and pseudo-angina pectoris have been described as pure vagus crises, but the interpretation is absolutely uncertain.

Between the crises and the lancinating pains stand the neuralgias, which are comparatively infrequent but obstinate. These *neuralgias* may attach themselves to the cerebral and peripheral nerves, in which case they resemble rheumatic neuralgias, or they may affect the internal organs. I observed for years a tabetic, who was tortured by the most violent unceasing enteralgia, which neither increased to the form of crises, nor ever assumed the type of lancinating pains.

To the subjective *sensory disturbances* of the later stages correspond more nearly, than in the early stage, *objective* changes of sensation, which we shall therefore speak of now. Even before the hyperæsthesia has completely risen in the legs, there generally appear zones of sensory disturbances in the trunk, which (*Hitzig, Laehr, Déjérine*) have segmentary arrangement (*cf. general diagnostics*), i. e., occupy the region of the skin, supplied by the individual posterior roots, resp. the segments belonging to them. Sometimes the existence of such zones is suggested by the girdle feeling of the patient (even in the region of the girdle pain the skin may be hyperæsthetic!), but often the girdle zones are absolutely independent, at the height of the breast or the costal arch, the umbilicus, etc.; very often, they are broader on one-half of the trunk than on the other, or are demonstrable only on one side, etc. The form of disturbance is here, usually a tactile hyperæsthesia or anæsthesia with a most evident hyperalgesia, also with diminution of the cold and heat sensations. Very remarkable also is the disturbance of the temperature sense in the trunk in the form of *hyperæsthesia to cold;* it appears usually early in the disease, often in the first stage, and the patient is also subjectively conscious of it. The tabetic becomes unduly susceptible to cold touches, especially to cold water, but also to other cold objects, etc. (usually less frequently to cool air!); such a touch almost causes him pain, he starts up, screams, etc. This hyperæsthesia to cold does not coincide with the girdle zones, which have just been mentioned, but generally affects the trunk from the costal arch downwards, often also the upper part of the leg, especially on its inner side. In these hyperæsthesias to cold a radicular (segmentary type) cannot be recognized. This disturbance is unusually characteristic for tabes, and is rarely absent even in the rudimentary forms.

Otherwise hyperæsthesias are scarcely ever met with in tabes. On the contrary in the later stage the loss of sensation to touch ascends the legs, passes over trunk and arms and may even attack face and head. The entire surface of the skin then, or only the surface of the body up to the neck, the breasts, etc., shows diminution of perception to a simple touch and usually

also to cold (with the one exception named), and heat. Total anæsthesia, even to coarser touch, appears only late, and confines itself mostly to isolated parts of the legs. On the other hand, *sensibility* to *pain* may be completely lost even at an early stage. Hypalgesia in feet and legs is found almost invariably. In addition to this, there appears very soon a characteristic *retardation* of the perception of pain stimuli. If the skin be pinched or pricked, the patient feels at first merely a touch, and only after several seconds does the feeling of pain follow, or the corresponding pain reflex, the withdrawal of the foot, etc. To this is often added a disturbance, which peculiarly supplements hypalgesia, the so-called *summation* of the pain stimulus. A stimulus of only short duration even though it be powerful calls forth no feeling of pain, but a *longer* pinching, or pressing the point of the needle against the skin, for a definite length of time does. In addition the feeling of pain often lasts for a considerable time afterwards. "*Abadie's* symptom," insensibility to pain when the Achilles tendon is pressed upon violently, is a very inconstant sign.

Concerning sensibility of the *internal organs* in tabes, we know but little. The normally great sensitiveness of the testicles and breasts to pressure disappears (*testicular analgesia*). The subjective feeling of the need of emptying the bowels and bladder, defecation and micturition, is disturbed early (cf. above).

Deep sensibility proper is always more or less influenced. The best known disturbance of this sort is the *Romberg phenomenon*, generally described as a disturbance of the muscle sense, which may be demonstrated in 90% of all tabetics; when standing with feet close together, and eyes shut, there is so pronounced a swaying of the entire body, that the patient may even fall to the ground, a phenomenon that is scarcely, or not at all, perceived, when the eyes are open, that is, when the patient can control his position with his own eyes. This symptom often belongs to the initial stage of tabes. But the disturbance is by no means sufficiently well explained by the otherwise unfortunate term "disturbance of the muscle sense" (Muskelsinnstœrung), for in it participates also the sense for passive movements, for gravity, for resistance, as well as finally for the actual perception of position.

But these sensory functions may be tested in other ways, too, and show, as has been said before, in tabes, as a rule, great changes in the advanced stages of the disease. The patients no longer feel how their limbs, toes, etc., are placed, bent, etc., by the examining physician—disturbance of the perception of passive movement; without the control of their eyes, they can not move their feet a definite distance apart—disturbance in perception of active movement; they do not know, where their legs are under the bedcover—loss of perception of position. In similar manner the senses for weight and resistance can be tested. Naturally, most of these tests prove the presence of *mixed disturbances* of the different sensory faculties. *Pall-*

æsthesia or the perception of vibration, which is connected with the sensitiveness of bones (*Egger*), is also, as a rule, much disturbed in tabes; according to some authors this proceeds parallel with the ataxia (see below), but according to *Egger's* latest report this does not seem to be the case. The *stereognostic* perception, finally, is disturbed more rarely, but occasionally in late stages it is very marked; the patients no longer recognize an object held in the hand, are no longer able to take money from their purses, etc.

The rather frequent absence in tabes of the *normal feeling of fatigue* after movements and exertions, is probably of other origin (*Frenkel*).

Partly, at least, as a disturbance of the sensory pathways, we may conceive the symptom we have not yet discussed and which is responsible for the name given tabes during its entire "second stage": *ataxia*. Even if there are cases of tabes absolutely without ataxia, this complete freedom from it is very rare; its presence in tabes is extraordinarily significant and characteristic, as nowhere else in human pathology do we meet it in exactly the same form and but rarely in a similar form. If the tabetic—in any position desired—is asked to perform complicated movements, especially with the extremities, there appears an uncertainty, a wavering; the movements wander from their direct line and proceed in zig-zag fashion. If the patient is to bring the tips of his index fingers together, by bringing the arms, which have been stretched far to the side, towards the front, this movement is attended by utterly unregulated waverings, extending on all sides about the axis of the movement. He invariably spills water from a full glass that he is to take to his lips; in writing, one letter becomes microscopically small while to form the next, the hand moves the pen all the way across the page. He is not able, when lying on his back, to perform the more delicate movements with his feet, to describe a circle with the points of his foot, to write numbers in the air, to place the heel of one foot, moving it through the air, upon the knee of the other leg. If he is asked to walk upon a line, he finds it very difficult, because his feet continually stray to one side or the other. On the whole the gait of the ataxic is peculiarly characteristic. At first it is swinging and stamping probably with the knee raised unnecessarily high and toes pointing downwards (like a rooster's walk). Later the legs spread apart, usually assisted by a cane, which is placed far in front, in order thus to attain as broad a base as possible for the three points of support. Even a slight degree of so-called *static ataxia* is occasionally present, in which the patient finds it less possible to sit or stand perfectly erect, without at the same time moving the muscles of the trunk, but far more frequent is the ataxia, which has just been discussed, the *locomotor* (or movement) ataxia, which in England and France has given the disease its name. All these atactic movements, moreover, occur even more typically when the eyes are closed, so that, to a certain extent, even the *Romberg* symptom, just mentioned, may be considered a partial phenomenon of ataxia.

Ataxia is to be understood as a disturbance of *co-ordination*, the main cause of which must be sought in a confusion of the centripetal stimuli—in the widest sense, therefore, of the sensory stimuli, though the disturbance, at first sight, appears to be a motor one.

Genuine, purely motor symptoms, however, are very infrequent even in advanced stages of tabes. In the greater number of grave cases of tabes, it is true, the patients are forced to stay in bed, and there is to a high degree, a feeling of general weakness, but this is altogether the result of sensory disturbances and ataxia. To some extent also trophic disturbances prevent motion. Paralyses are known almost only in the area supplied by the cerebral nerves, if we leave out of consideration the rare combinations with *atrophic paralyses*, described by *Déjérine* and others, which are probably partly dependent upon implication of the peripheral motor nerves, partly the result of disease of the gray anterior horns, and which, therefore, in neither case, belong to the picture of tabes proper. In these cases, we find conditions of weakness and atrophy, especially in the small muscles of the feet and hands, in the area of the peroneal, as well as in those of the ulnar and median nerves; secondary contractures, the pied bot tabétique (tabetic club foot), claw and monkey hand are the consequences. The reaction of degeneration is not demonstrable in the majority of the cases. But the general emaciation, so frequently observed in the terminal stage of tabes, is only rarely due to these changes, much more frequently it depends upon disturbances in nutrition, and disuse of the muscles (marantic tabes). More frequent are, as has been mentioned, the paralyses of individual *cerebral nerves*.

The *olfactory* is affected only in very rare instances.

The disturbances of the *optic* nerve—amblyopia, diminution of the visual field, white atrophy—have been mentioned; they are not at all common.

The *oculomotor*, *trochlear* and *abducent* nerves are more frequently and also to a more considerable degree affected than the other nerves; this also has been mentioned above. Outside of the paralyses of individual nerves and the ophthalmoplegias, there occur also accommodation paralyses—scarcely ever isolated—as well as, but rarely, irritation phenomena (convergence spasm) which like the club foot in tabes may also be considered as *paralytic* contractures—possibly with better reason. The appearance, however, of "*disturbing*" double images, is by no means very frequent, in even the serious forms of tabetic paralyses of the ocular muscles, since the patients are often able to suppress them.

Disease of the *trifacial* causes frequently, in advanced cases, more or less severe disturbances of the cutaneous sensibility in the face, particularly in the conjunctival mucous membrane, which especially when associated with lagophthalmos (this is, of course, rare) may lead to the formation of ulcers,

and ophthalmia. The radicular (segmental) type may be recognized here also. Very rarely the motor trifacial is affected causing weakness in the muscles of mastication.

In the *facial*, paralyses as well as spasms (tic convulsif) have curiously enough been described very infrequently, but both occur.

The *auditory*, again, is more frequently affected and labyrinthine vertigo as well as disturbances in hearing are described, alone and together, that is, disturbances that include part, or the whole of the *Ménière* symptom complex. In the largest statistical paper (*Tumpowsky* in 225 cases of tabes), a proportion of 0.9% of affections of the auditory nerve was given, a figure which probably is too small, and could certainly be raised by exact otological examination.

Glosso-pharyngeal Vagus.—Of taste disturbances all varieties are reported, though, on the whole, they are of rare occurrence. Absolute ageusia, loss of the sense of taste, as well as confusion of the separate qualities of taste, occurs. Severely affected tabetics frequently say that everything tastes to them "like wood"; with closer investigation, however, there is proved to be only a partial lowering of the separate qualities of taste. One observation of *Pfeifer* mentions, on the analogy of the delayed pain sensation, a retarded taste sensation. This group of phenomena, as a whole, belongs to the domain of atypical forms of tabes, especially to those of the bulbar-paralytic type.

A pure vagus symptom is the not infrequent *tachycardia* of tabetics; at rest, the pulse has been observed as high as 120. Tachypnœa may belong here also. The disturbances of the laryngeus inferior—seldom permanent paralyses, most frequently posticus paralysis, occasionally spasms, now and then the paroxysmal pareses of the laryngeal crises—lead us to the crises that belong almost essentially to the vagus group, and which were discussed above. Here we must emphasize the fact, that, if we wish to consider the gastric crises, etc., as vagus symptoms, we must assume in numerous cases an early vagus affection almost parallel to that of the opticus. I believe, therefore, as has been mentioned already, that part of the crises must certainly be due to *affections of the sympathetic nerve.*

Spinal Accessory Symptoms.—Pareses of the cucullaris and the sterno-cleido-mastoid have recently been described by Seiffer; they must belong, at any rate, to the rarities, as do also the affections of the *hypoglossus*, which occur occasionally accompanied by atrophy.

One unusually frequent motor symptom, the loss of *muscle tone*, we have not as yet spoken of. The reflex inhibitions, normally present in every active muscular innervation, which prevent the movement from going beyond its goal are often lost early in tabes (nearly always in the final stages). Therefore the purposive movements, when the strength is normal (even when ataxia is not present), make an excessive impression, which is specially

increased by every passive movement. Without any trouble and without any sensation of pain the legs of the patient may be flexed to such a degree as to touch the head; the hyperextensibility of the fingers and wrist often makes it possible to bring the fingers passively into contact with the dorsal surface of the forearm; the leg extended at the knee, with the patient standing upright, is bent backward by the weight of the body (genu recurvatum). At the same time the muscles, ligaments, joints are anatomically, as a rule, perfectly normal. (Of course trophic changes in the joints may greatly increase the extent of passive movement.)

Electrical changes in the muscles are naturally to be expected only when peripheral paralyses exist and have been so reported.

Of the *motor irritation phenomena, athetosis* is a not very infrequent symptom of irritation in advanced tabes, in tabes superior, and in the bulbar-paralytic form; it gives the complete, uni- or bilateral, clearly defined picture of the movement disturbances just as they are observed in other cerebral diseases: continuous, slow, worm-like movements, which may lead to apparent dislocations, to overextensions and to quite grotesque positions, especially in the fingers and arms. *Contractures* and their concomitant symptoms—exaggeration of the reflexes, *Babinski* symptom, etc.—are observed only in the combination of tabes with other spinal troubles. That the *crises*, especially the laryngeal, represent partly paralyses, partly also spasmodic spastic conditions, has been said above.

Genuine, pure *sympathetic symptoms*—widening of the palpebral fissure, unilateral blushing and sweating, etc.—have been found only exceptionally in tabes.

The change of the *tendon reflexes*, which characterizes even the initial stage of tabes, is practically constant in the ataxic stage. The Achilles and patellar tendon reflexes are those most frequently absent; the patellar reflex can only very rarely be evoked even by artificial reenforcement (*Jendrassik*). The triceps and the lower jaw reflex are constantly absent in tabes superior but are not necessarily absent even in advanced cases of the usual type, beginning with the legs, since the reflex arcs for the arms and the head may remain undisturbed for a long time. Occasionally, however, the tendon reflexes that have been lost, may return temporarily or permanently, without a cure being effected, as in slight narcosis, after cerebral apoplexies, and upon the advent of other systemic diseases. Occasionally the reappearance seems, symptomatically, to show a certain improvement, especially when there was originally only a difference in the reflexes on the two sides, and then, one disappeared to return again. This, at any rate, does not seem to be such a very rare occurrence. In patients, moreover, in whom the tabes, from the beginning, appeared in the form of a *combined* systemic disease, a tabo-paralysis, etc., the reflexes can be retained from the first, or even increased.

The *cutaneous reflexes* are altered very much more rarely, most frequently the plantar. This is lost in most tabetics, as soon as the sensory disturbances in the feet have attained a certain degree; their decrease is parallel, on the whole, with the loss of feeling (which by no means holds good for the tendon reflexes). The so-called Babinski dorsal toe reflex is never present in pure tabes. The cremaster reflexes, and especially the abdominal reflexes are, on the other hand, only exceptionally affected by the tabetic process, even where zones of anæsthesia exist in the trunk. Whether the activity of the abdominal reflexes, which is not rare, is connected with the hyperæsthesia to cold in that region, has not yet been ascertained.

Barring the pupillary reflex already mentioned, the so-called "*internal reflexes*" (of swallowing, sneezing, etc.), are disturbed only exceptionally, unless we are willing to accept some of the crises, as the laryngeal crises, etc., as due to reflex overexcitability, a theory which certainly would not hold good for the majority of cases. Formerly several forms of male impotence were explained as due to reflex disturbance (faulty ejaculation).

The symptoms referable to the *urogenital tract* in tabes, on the whole, present a mixture of motor, sensory, and reflex disturbances; they have already been discussed in part with the early symptoms.

In the completely developed clinical picture of the disease, they are rarely ever altogether absent, and the bladder disturbances especially, very frequently are of great importance in the termination of the disease, causing death by ascending pyelonephritis.

The *bladder disturbances* are also the most frequent disturbances in this group. The patients must in many cases use a greater amount of strength to empty the bladder, and there usually remains some residual urine—retentio urinæ; this is mostly caused by spasm of the sphincter, but occasionally there is also simultaneous weakness of the detrusor. This, however, is not constant; it may even happen in tabes, that, because of lasting spasm of the sphincter, a genuine hypertrophy of the bladder, a so-called "board-bladder" (Balkenblase) may develop with partial hypertrophy of the muscle of the detrusor. The retention may lead to the clinical picture of ischuria-paradoxa: the constantly filled bladder, unable to empty itself, emits passively, so to say, small quantities continuously, like an overflowing rain-barrel. Naturally such patients very often take shelter in the last resort—the catheter; usually the bladder becomes infected from this, sooner or later, so that cystitis in grave cases of tabes is often found as a direct result of this. This, however, is also caused by the opposite condition to retention; namely, incontinence of urine—the relaxation of the sphincter with constant dripping of urine. In this condition also, which is almost as frequent as retention, infectious micro-organisms easily find their way through the urethra into the insufficiently closed off bladder. Disturbances of the *intestinal sphincters* are found less frequently; futhermore all proof for the statement

that the constipation, which is as frequent in tabes as in other diseases of the central nervous system, is induced by a spasm of the sphincter is lacking. It is easier to believe that it is occasionally caused by a chronic paresis of the intestinal muscles, or a genuine ataxia, which, however, frequently seems to affect the sphincters alone. Often patients are troubled by the lack of control over the emission of urine or of evacuation of feces, due to a deeply seated anæsthesia.

Impotence is one of the most frequent tabetic symptoms of this group, known, as such, even to the layman. Almost all male patients—in women disturbances of libido and orgasm are seldom even observed by the patients themselves—show, in advanced stages of tabes, a decline in virile power, which affects all the factors of this function; erection and ejaculation in particular become insufficient, take place prematurely, or are even painful, whereas the libido, to the torture of the patient, may last for some time after the loss of potency. In the final stage there is generally total impotence. In the beginning of the disease, on the other hand, the presence of *satyriasis* may be noted.

Of the ordinary groups of tabetic symptoms, only the trophic disturbances and the changes in the cerebro-spinal fluid remain to be mentioned.

The *trophic disturbances* may affect all the tissues of the body, but most characteristically, the epidermis and the skeletal system. The deficient nourishment of the skin is betokened frequently by ulcers and bed-sores; wherever a tight bandage, a set of false teeth, a urine bottle have pressed for some time, ulcers appear, which occasionally become very deep, have but a slight tendency to heal and are nearly always painless. Such an ulcer, which appears in tabetics apparently spontaneously, is the "*mal perforant*," usually coming on gradually on the under surface of the great toe, or of the ball of the foot back of the great toe, which, as a rule, eats in very deeply, even to the bone, and is difficult to heal. Also an (apparently) spontaneous herpes zoster is often observed in tabetics. Still more striking are the trophic disturbances in the bones and joints, more particularly the *arthropathies* (Fig. 60). Suddenly, without any spontaneous pains, and within a few days, any joint of the body may become swollen; the patient, as a rule, does not notice it until it causes purely mechanical disturbances in motion. Upon objective examination, however, one generally finds serious changes; often an enormous distention at the ends of the bones and effusion into the joint. Local heat and redness, as well as pain, are absent. An X-ray examination shows an indistinct clearing up of the shadows of the bones, which, however, retain their sharp contours; the anatomic examination often shows detachment of the periosteum with softening of the parts of the bone beneath, formation of osteophytes, disintegration of small particles of bone, destruction of ligaments and joint cartilage. The exudate is, as a rule,

serous and but rarely contains pus. The consequences are loose joints, subluxations, etc.; because of the chronic course and the very slight tendency to heal, permanent deformities often supervene, which, combined with other tabetic symptoms, especially with hypotonia of muscles, may lead to almost grotesque pictures of diseases, as, for instance, to the not infrequent genu recurvatum with synchronous arthropathies. As a matter of fact, the arthropathies are found most frequently in the knee-joints, then come the

FIG. 60.—Arthropathy of the right knee in tabes dorsalis.

hip-, the shoulder-, the ankle-joints, etc. In more recent times, their occurrence in the spinal column has been described with striking frequency; probably the general employment of radioscopy may have contributed to this. To a certain extent the *abnormal fragility of the bones* in tabes, runs parallel with the arthropathies; even slight exertion leads to complete, though painless transverse fractures of the long bones, which, however, show a greater tendency to heal than the arthropathies. Dentists, moreover, frequently observe a strikingly rapid deformation of the jaws, especially of the alveolar processes, with falling out of the teeth; the process resembles absolutely the

atrophy of the senile jaw, but is more rapid and appears earlier. The change often takes place so quickly that sets of false teeth, made to fit the mouth, no longer fit properly at the end of a few months.

The last symptom which must be included among those that are almost constant in tabes, is the *change in the cerebro-spinal fluid.* If a few cubic centimeters of the liquid are withdrawn, in the usual way, by lumbar puncture, from the spinal canal of a tabetic, and subjected to centrifugalization, there is regularly found in the sediment an increase of the normally rather rare cellular elements, and especially of the lymphocytes, so called because of their likeness to the lymphocytes of the blood; there is usually a very high degree of "lymphocytosis" of the fluid (*Widal, Sicard, Schoenborn, Nissl*). A similar change is found in pure meningitis, as well as in some other metasyphilitic diseases of the central nervous system—progressive paralysis, cerebro-spinal syphilis. The most likely interpretation of this symptom in tabes is a chronic change in the meninges brought about by the former attack of syphilis—a syphilosis of the meninges. The supposition, that the former attack of syphilis in itself causes the lymphocytosis in tabetics—in a certain number of cases similar findings have been made in secondary syphilis—becomes improbable, since upon closer investigation of the meninges in tabes, anatomic changes are now almost regularly found (*Nageotte*, cf. above). Lymphocytosis is an unusually constant and early symptom of tabes dorsalis.

There are several groups of symptoms, which *combine* now and then with those of tabes dorsalis. Among these are the symptoms caused by the participation of those systems of tracts or columns in the spinal cord which, as a rule, are not affected in tabes, especially that of the pyramidal tracts. Then we have before us the complete picture of "combined system disease," usually accompanied by spasticity and exaggerated reflexes. The combination with peripheral neuritis, also, is not infrequent. The most frequent concomitant disease of the nervous system is probably *progressive paralysis.* But this, as *Erb* pertinently emphasizes, must not be interpreted, as if tabes, when reaching a certain high level, very frequently passed over into progressive paralysis; this, on the contrary, is a relatively rare occurrence. The combination is, in the first place, rather an anatomical one—in autopsies of paretics a gray degeneration of the posterior columns (*Westphal*) is often found, and since then it has been learned that occasional tabetic symptoms (Romberg, pupillary rigidity, slight disturbances of sensation) may very often be found in paretics upon closer investigation. Whether, in such a case, the tabes was superimposed on a paresis already present, or whether the two developed simultaneously, we are generally unable to decide; either might happen. But *tabo-paresis*, though a frequent disease picture, is not nearly as common as simple paresis, and especially not nearly so frequent as simple tabes. Otherwise, psychic symptoms in tabes are rare, but

combinations with so-called "neuroses," epilepsy or Basedow's disease may occasionally be observed.

We have already stated that tertiary syphilis may occur with tabes. But in particular we see certain late forms of visceral syphilis in tabes, especially *aneurism of the aorta*, to such an extent, that *Babinski* has described a peculiar symptom complex: aneurism of the aorta, with disturbances of the tendon and pupillary reflexes. In the great majority of cases, the aneurism, as well as the not infrequent insufficiency of the aortic valves, might be caused by a syphilitic aortitis.

The occurrence of chronic gastro-intestinal disturbances and ascending pyelonephritis in tabes, is explained by the foundation for these troubles by the tabetic disturbances of the respective sphincters.

How far, then, can a **pathogenesis** of the tabetic **symptoms** be established, how far can they be traced back to the anatomic process? This question is easily answered in regard to some phenomena; for others up to this time with great difficulty, or not at all. The objective, and to an extent, the subjective *sensory disturbances* are founded, doubtless, upon a degeneration of the posterior root fibres; this is proved in many cases by the complete radicular distribution, and the ascent of the disturbances, etc. But even here, much is left unexplained. Thus, for instance, the "sensory disturbances of the internal organs," especially of the intestines and bladder, seem to be directly or indirectly connected with degeneration of *sympathetic* tracts. Also the reason why so frequently certain qualities of sensation are disturbed more seriously or earlier than others, has not been definitely established thus far. The explanation of the tabetic *pains*, especially of the *lancinating* pains, and of the so-called *crises*, is most difficult. For the former, which show a predilection to limit themselves to the area of a peripheral nerve, it is possible to consider irritation or compression of a circumscribed place of the posterior roots, even though the type of pains, which certainly depends on such compression (in pachymeningitis, tumors, herpes zoster), resemble but slightly, as a rule, the lancinating pains. Finally as to the crises, we know, at present, no anatomic basis. Their localization, their character, their connection with considerable increases in blood pressure, tempt us very much to refer them to the vasomotor nerves, i. e., to the sympathetic nerve system.

The *disturbance of the muscle sense*, *Romberg's* phenomenon, probably must be traced back also to a disturbance of the centripetal paths, most likely of the fibres ascending in the posterior column; pallæsthesia and its disturbance belong here also, and for the stereognostic disturbances we must assume, in contradistinction to the astereognosia of cerebral affections, a mixture of centripetal and centrifugal, sensory and motor disturbances. On the other hand, the frequent absence of the feeling of fatigue in tabetics,

represents certainly a pure disturbance in the sensory rapport of the extremities, that is, in the posterior columns.

The understanding of *tabetic ataxia* has long caused great difficulties. If we consider briefly its manifestations, we shall see immediately that a motor disturbance is out of the question. The patient is able to properly innervate every single muscle, at least with the aid of the control of the eye; the impulse of the will to the periphery, i. e., the motor conduction is therefore absolutely retained in the usual forms of tabetic ataxia. What is disturbed is, first, the sensory rapport at the periphery, the "information" of the central organ as to the position of the separate divisions of limbs, and skeletal muscles, but then again, the synergism of the individual muscles, especially the relation between agonist and antagonist. For the lack of sensory rapport, we have the anatomic basis: the degeneration of the posterior columns (in the broadest sense); for the co-ordinations, we should have to consider, the so-called co-ordination centers in the cerebellum, pons, quadrigeminal bodies; they are certainly not affected, for the most part, in tabetic ataxia. But co-ordination, also, needs the centripetal, sensible (sensory) impulses; it is natural, therefore, to assume here, too, a genuine disturbance of the centripetal paths. Without entering here upon the moot points in detail (which, in Erb's opinion, are not, as yet, fully cleared up), let us mention only that hypotonia of the muscles (cf. below) as well as reflex processes seem to play some part in tabetic ataxia. But in the main, it is doubtless a question of a "*sensory*" *ataxia* (*Leyden*) and of a disturbance of the sensory paths (*Otfried Förster*). The last named author sees the anatomic basis of tabetic ataxia in the degeneration of the reflex collaterals in the spinal cord, the collaterals to Clarke's columns (cerebellum) and the long fibres of the posterior column. I fully agree with his point of view.

The disturbance of the tendon reflexes is certainly founded upon the degeneration of the reflex collaterals, of the root fibres passing from the radicular zone into the gray posterior columns. (Level for the patellar reflexes: lumbar segments II to IV; Achilles tendon reflexes: sacral segments III to V; triceps reflex: cervical segment VI–VII.) Hypotonia, also, perhaps even the disturbances of the *cutaneous reflexes* (rare) are to be traced back to the reflex collaterals. For the "*internal reflexes*" and the disturbances of the *urogenital tract* (the so-called sphincter disturbances, incontinence and retention of feces and urine, impotence), the genesis, as we mentioned above, is not absolutely clear. We know, to be sure, that the lumbar segments are the seat of the corresponding centers. But animal experiments seem to indicate that the path for the processes mentioned passes at least in part through the sympathetic, for the co-affection of which in tabes dorsalis unfortunately only a limited number of reports are available.

The *motor centers* and paths need occupy us less. In the rare atrophic paralyses, corresponding lesions of the anterior horn have been found, and here and there also neuritic changes. Of the irritation phenomena, *athetosis*, according to the present state of our knowledge, is certainly a cerebral symptom. In the phenomena of the *cranial nerves* (nerves of the eye muscles, special sense nerves, trigeminus, vagus) there appear with about the same frequency, neuritic and nuclear lesions; optic atrophy especially is, as stated above, before all a sign of a peripheral affection.

The *changes in the pupils*, especially the reflex light rigidity, have always given much concern to neurologists and ophthalmologists. Experimental investigations (Bach and H. Meyer) seemed to prove that in the posterior columns, near the fourth ventricle, that is in the uppermost cervical segment, there is a center for the pupillary reflex (and for the origin of so-called "spinal miosis"); but recently Bumke has found proof to the contrary, and now it almosts seems, as if one should have to return to the older theories, according to which, pupillary rigidity is to be viewed as a lesion in the pupillary fibres of the optic nerve, in the nuclei of the oculomotor or in the ciliary ganglion. But no certain results have been established for the localization of this apparently simple symptom (i. e., for the anatomic proof).

For the *trophic* disturbances, to be brief, we have, at present, absolutely no satisfactory localization in the central nervous system; only this is certain, that they are not to be understood only as local, idiopathic destructive processes of the bones, etc. The changes of the *cerebro-spinal fluid*, finally, are to be explained only by the assumption of a chronic spinal meningitis, perhaps starting especially from the posterior roots.

The course and prognosis of tabes dorsalis, present on the whole, and in spite of some individual differences, a very sorry spectacle. The preceding observations show the *usual course*. The patients mostly suffer for a few years from lancinating pains, perhaps also from slight disturbances of the bladder. With sufficient self-observation, they recognize already a slight disturbance of sensation in the feet, some uncertainty in walking in the dark; the examination at this time usually discloses pupillary changes, and a difference in or absence of the Achilles and patellar reflexes, possibly also some hyperæsthesia. Diplopia, disturbances in the muscles of the eye, impotence also appear. As a rule years, ten or more, have passed since the first "lightning pains." Then are added increased disturbances of sensation and ataxia in the legs, gradually also in the arms, which robs the patients more and more of the ability to move (the demarcation of a third stage in the symptomatic picture of tabes, the paraplegic stage, mostly resting upon this, I consider as arbitrary). The further extension of the process to the cortex of the brain, is usually less clear, since some phenomena of the cerebral nerves appear much earlier; on the other hand, certain cerebral symptoms never or very rarely (athetosis) supervene, and the transition to paresis, as

has been mentioned, is not frequent in typical tabes. The final issue is usually fatal, though death relatively infrequently is due to direct symptoms of the disease (inanition in gastric crises, posticus paralysis); more frequently it is due to intercurrent diseases (infective diseases) and especially to secondary consequence of tabetic processes: sepsis due to trophic disturbances, bed sores or cystopyelonephritis, as the result of external infection due to the sphincteric lesions.

There are several characteristic variations from this usual type. In the first place, the course may be very much more rapid, and it is generally the first stage which is very much shortened; in such instances the first paroxysms of pain may be followed by ataxia in a few weeks, while the later stages may extend over the usual long period of time. Other characteristically atypical forms are *juvenile tabes*, appearing in children, almost without exception, upon a congenital syphilitic basis, and, as a rule, of a relatively benign character (thus far only a single autopsy of an indubitable case has been reported), and tabes superior, which, on the other hand, is a very unfavorable form of the disease. In *tabes superior* the symptoms begin in the upper extremities, with pain in the ulnar area and paræsthesias, also triceps reflex disturbance, whereas the tendon reflexes in the legs may be retained; in such cases bulbar symptoms, dangerous to life, usually supervene very early.

In addition to these types, authors who have studied a large number of tabetic patients, like *Erb* and *Déjérine*, have distinguished additional clinical *types*, according to the predominance of one or the other symptom: tabes dolorosa with especially violent pains; tabes visceralis with violent crises and without specially marked subjective concomitant symptoms. Even though the value of this classification can be practical only in so far, as such cases usually preserve their characteristic form for the entire and usually protracted course of the disease, for *one* form the predominance of one symptom is so significant, that a particular anatomic basis has long been looked for: tabes with *early optic atrophy*. According to the investigations, especially of the school of *Pierre Marie* (*Lêri and others*) 25% of tabetics lose their sight in this way; in these cases the rest of the nervous system, frequently participates with cerebral symptoms, but rarely with spinal symptoms of marked gravity (with the exception of a regular loss of the tendon reflexes); the further course also, usually protracted, corresponds to this picture so that a few authors interpret "tabetic amaurosis" and the usual form of tabes as two different localizations of the same process. Finally to the deviating forms of tabes, there belong also the cases of *imperfectly developed, rudimentary tabes*, of which much has been published in recent years. The conception is not easily defined; as a matter of fact, one can speak of rudimentary or "abortive" tabes (*Möbius*) only when the condition, once discovered, persists stationary throughout or at least remains

thus for a very long time, and, finally, shows only *few* symptoms of the disease. The possible combinations of such individual symptoms are, as Erb emphasizes, naturally very numerous and varied, but in almost all the cases, there is found one of three symptoms: loss of tendon reflexes (if only of one single Achilles reflex); pupillary changes (possibly only miosis without loss of the reaction to light); lymphocytosis of the cerebro-spinal fluid. Naturally in such a patient, whose affection is recognized accidentally, possibly as he is being examined by the physician because of "rheumatic pains," one cannot tell at once, whether he will not end by becoming afflicted with a fully developed tabes, but not rarely this is not the case, and we may then assume without further investigation, that it is a question of fixed, quasi "checked" pathological changes, which have possibly affected only the posterior roots and meninges and did not go any further than the entrance into the spinal cord. In these cases, the addition of even *one* new symptom during the period of observation is usually the signal for a further development of the disease, which then generally continues steadily to progress. Naturally, this rudimentary tabes, is an especially interesting field for diagnosis.

The **prognosis** of tabes is apparent, from what has been already said. It is, quoad sanationem, *bad*. Cases of genuine recovery, i. e., of complete disappearance of the subjective, and a total or almost total disappearance of the objective symptoms (areflexia of the pupils or some similar symptom alone remaining), as shown in the pathological changes brought to light by post-mortem investigations, are exceedingly rare, but do occur occasionally. Even the disappearance of single symptoms is rare; in regard to this *v. Malaisé* (from *Oppenheim's* polyclinic) recently reported the following: The *lancinating* pains improved in 37% of the patients (in forms that ran a wholly favorable course), the *bladder disturbances* in 8 to 10%, the sexual power never, when once disturbed (but here one must consider the physiological decrease that normally appears with advancing years). Very frequently, on the other hand, the crises that have once appeared, disappear, that is, they do not return, whereas *atrophy of the optic nerve* nearly always leads to total blindness, occasionally only after ten years, on an average after 5 1/2 years. *Ataxia*, as we shall see, can be improved by therapeutic measures, and perhaps even spontaneous improvement is possible; in regard to the *paralysis of the muscles of the eyes* we have mentioned already, that even in their intensity, they are usually subject to extraordinary fluctuations.

Cases, in which the so-called late symptoms appear early, are, as in most diseases, prognostically unfavorable (ataxia, trophic disturbances); the longest possible extension of the first stage is rightly considered favorable. Frequently, also, the disease remains stationary for years, and under favorable conditions of life, the patients may live to a very old age, so that according to French writers (*Bellugaud and Faure*) about 59% of all tabetics

experience an "evolution benigne," 36% an "evolution grave," and 5% recover. I consider these statistics too favorable, and should prefer to subscribe to the statement of *v. Malaisé*, who out of 70 patients, found 2 completely freed of their troubles, 26 retaining their working capacity for a long time, and in good general condition, 30 with constant but slow progression and 18 running a rapid and unfavorable course. Some factors mentioned when discussing the ætiology, influence also the further course of the disease. Especially unfavorable results come from alcohol, sexual excesses, traumatisms and physical over-exertion, extreme degrees of temperature, persisting cares and excitements.

Somewhat more favorable is the question of *prognosis quoad vitam. Pierre Marie* assumes that the length of life is not shortened by tabes (51.5% died after the sixtieth year) and even with the less favorable material of *Oppenheim*, the statement that the majority of the patients die after the sixtieth year, holds good.

The diagnosis of tabes, under ordinary circumstances, is easy for everyone who understands how to make a neurological examination. The unfortunately still very frequently occurring errors in the diagnosis of the disease are to be explained only by a lack of such knowledge. The above mentioned abortive cases and occasional cases with peculiarly complicated symptoms or course, are the only ones which may cause the neurologist difficulty. We have mentioned several times already the cardinal symptoms, of which one, at least, is to be found also in the abortive tabes, the "tabes fruste": *disturbance of the tendon reflex, pupillary disturbance, lymphocytosis* of the cerebro-spinal fluid. If all three are absent, we may, with a considerable degree of certainty, exclude tabes; at the most, patients with persistent pains, like the lancinating ones, should always arouse a suspicion of tabes, which, however, would have to be fully confirmed sooner or later by the appearance of one of the symptoms named. Each of these cardinal symptoms, that may accidentally be found in a patient, must, in itself, suggest the possibility of the existence of tabes; if, in the anamnesis syphilis appears, or if, in the history of the existing disease, possibly a paralysis of the muscles of the eye is spoken of, or if, in the objective examination, hyperæsthesia to cold in the trunk is found, the thought is so much more reenforced. Naturally some of these phenomena appear also in other diseases, with the exception, perhaps, of the *hyperæsthesia to cold;* at any rate, the combination of some such symptoms, as well as the further course of the disease, usually justify the diagnosis of tabes.

Of the abortive cases we have already spoken. If we see, for instance, a patient with slight pupillary differences, lancinating pains, and owning to a history of syphilis, I recommend always, beside the most careful examination as to sensation and tendon reflexes, above all to perform a lumbar puncture for purposes of diagnosis; if this turns out negatively, I am inclined to exclude

tabes or at least consider it most improbable; a positive result in lumbar puncture makes the diagnosis of a metasyphilitic disease of the central nervous system (according to its frequency, probably a tabes) almost certain, in so far as, and this is easy for the most part to determine, another meningeal process can be excluded. I should arrange the diagnostic symptoms, in the order of descending importance, as follows:

1. Loss of the tendon reflexes.
   Miosis and reflex rigidity of the pupils to light.
   Lymphocytosis of the cerebro-spinal fluid.
2. Hyperæsthesia to cold.
   Other sensory disturbances of tabetic type, especially, however, summation and retardation in conduction of pain.
   Anamnesis: lancinating pains; former syphilis.
3. Paralyses of the muscles of the eye.
4. Romberg's symptom.
   Ataxia.
   Hypotonia of the musculature.
   Sphincter disturbances, impotence.
5. Optic atrophy.
   Crises.
   Trophic disturbances.

For the *great variety* of symptoms to produce diagnostic difficulties, is a relatively rare occurrence, and happens almost only when totally foreign phenomena are present in addition to the complete picture of tabes; for instance, hypertension of the muscles, exaggerated tendon reflexes, peripheral paralyses. As a rule, then, it will be a question of combination with other diseases—sclerosis of the lateral columns, peripheral neuritis—only exceptionally of a pure tabes with an atypical course.

Excluding the combinations we have just mentioned, especially the so-called combined systemic diseases, which vary in their symptoms according to the greater participation of the posterior or the lateral columns, only the various forms of *neuritis* are of *differential diagnostic* importance, if we except also the rare, usually epidemic ergotin poisoning (ergotin tabes), the symptoms of which correspond almost exactly with those of real tabes. Of neuritic processes, alcoholic neuritis most frequently hampers our diagnosis, more rarely the diphtheritic and other toxic forms of neuritis (diabetes mellitus with neuritis). Whereas in these patients, the loss of the tendon reflexes and the disturbances in sensation may remind one strongly of tabes, the motor paralyses, and, above all, the promptness of the pupillary reaction, lead usually to the correct diagnosis. Multiple sclerosis and syringomyelia may also upon occasion, enter into the problem of the differential diagnosis.

In spite of the bad prognosis, the *treatment of tabes*, corresponding to the chronic character of the disease, is even to-day uncommonly diverse, and,

let us add, by no means devoid of results from the standpoint of alleviating the suffering of the tabetic.

Treatment must be aimed at two points, inasmuch as only in the rarest cases, is there hope for complete recovery. It must be adapted to the existing stage of the disease, and to the degree of the result that probably may be attained, and must not, by the violence of applications, or by meddlesomeness physically or mentally over-exert and fatigue the patient.

Because of the meta-syphilitic nature of the disease the first thing to do, is to try a course of anti-syphilitic treatment, to give first mercury or iodides. Owing to the differing views of various physicians and "schools" this plunges us at once into a region of dispute. There is comparative agreement as to the fact, that in particular mercurial treatment does as a rule no harm, and in some few cases produces a slight, but definite beneficial effect. From this middle point, however, the positions of the extreme camps deviate considerably in both directions. While some physicians reject the Hg. cure as useless and occasionally injurious, Erb, especially, defends it earnestly; in his experience, it does "absolutely no harm," is well tolerated, and has often an excellent influence especially on the lighter symptoms of the disease (disturbances in sensation, etc.). I, myself, should like to say that I never saw mercurial treatment make the condition worse (I am a little doubtful only in cases of patients with optic atrophy, though I can *not* confirm the harmful influences of the treatment described by some authors). Direct favorable consequences, which could be ascribed *only* to the Hg. cure, I have seen but rarely; on the other hand, it has been quite the usual thing in my cases, that with regular treatment by inunction, repeated annually or every other year, the patients were subjectively in good condition, were able to perform work, gained weight, and remained for years about the same. Some of the patients ascribed considerable improvement to the inunctions and asked for them again and again. Naturally, even mercury fails to produce results in a number of cases, and I have not succeeded definitely, any more than have most of the French authors, in discovering in the lymphocytic content of the spinal fluid—which denotes the condition of the meninges—any difference worthy of mention before and after the treatment. On the whole, I *recommend* mercurial treatment in recent cases (it constitutes, in my opinion, the only *prophylactic* method worthy of recommendation, after syphilis has been acquired, especially for neuropathic individuals, though one must concede, that even after syphilis has been treated with the most painstaking correctness, a tabes may appear later), in syphilis still active in association with tabes, and in general in those cases in which, after definitely or probably diagnosed syphilis, no mercurial treatment has been used for some time. The only condition is that of moderate vigor of the patient and the possibility of control of the treatment by the physician. If syphilis may be excluded with probability, or many courses of treatments have already proved ineffec-

tual, I forego mercury as a rule. Concerning the method of application, I have like Erb, had the best results from small, frequently repeated inunctions (daily, for 30 days, 1 to 2 ℥ mercurial ointment); the usual care of the gums for the prevention of stomatitis is especially important. Concerning the use of iodides, there is less difference of opinion; in addition to or between the Hg. treatments, I administer for a space of 3 to 6 weeks, iodide of potassium (15 to 30 grs. pro die), also iodipin (subcutaneously) or saiodin (20 to 30 grs. pro die), and have seen from this treatment only favorable results; special consideration must be given the appetite and digestion of the patient, which therefore often requires that internal medication be replaced by subcutaneous administration of iodipin, which otherwise is not agreeable to either patient or physician. Serological syphilitic therapy unfortunately has, up to the present, shown no effect on tabes. Now the entire host of nervina follows; in order to make a change here and there, the physician must have at his disposal a sufficient supply of remedies. *Ergotin* (secale cornutum 0.5 pro die) I have used but seldom; *Charcot's* school, however, formerly recommended it warmly. *Argentum nitr.* I give occasionally, preferably in pills (for instance: Arg. nitr. 15 grs. strychni grs. ii. in 100 pills, one daily three times), for several weeks, in combination or in alternation with preparations of *strychnine*, of which, especially in the form of Erb's "tonic pills" I have seen excellent results (Ferr. lactic. 25 grs., extr. chin. aq. 25 grs., Extr. nuc. vom. 7 to 10 grs., extr. gentian. q. s. to 100 pills, take six daily). The arsenical preparations in their pure form I do not like very much, on the other hand, I can recommend warmly the cacodyl preparations (natr. cacodyl. 0.02–0.1 subcutaneously pro die) from my own experience, for continual treatment. Of atoxyl, which has recently been used as an anti-syphilitic agent, I have seen little benefit in tabes. Of general tonics, one must mention in addition, and for occasional trials, glycerophosphate, lecithin, nucleogen, thiodine, recently inaugurated by the French, phytin, etc. The same purpose, preservation of strength, beside the nourishment of the patient is served by the host of medicines that rouse the appetite, and the artificial food preparations. (Salvarsan is of no benefit in tabes, in my experience, and may do harm. Burr.)

As an attempt at local treatment at the place of the disease, we may name here the various methods of influencing the spinal cord and the posterior roots. In the front rank stands the *galvanic* current, which may be used in almost every stage and with almost every patient.

To be recommended is the stabile use of the constant current; two equally large, medium sized electrodes are used, one on the nape of the neck and the other over the lumbar enlargement, using a current of 8 to 10 milliamperes, alternately 3 to 4 minutes in each direction, without brusque interruptions. Frequently also I apply a larger electrode to the spine, a smaller to an extremity (peroneus point, wrist) or use Erb's sympathetic galvanization; the small cathode over the cervical sympathetic, the larger anode over the

opposite half of the body, placed in three "stations" near the spinal column, using a current of about equal strength. Symptomatically, also, especially in enteralgias and weakness of the bladder, the galvanic current is valuable: medium sized electrode over the lumbar segment of the cord, a large one over the abdomen or bladder. I use the faradic current (medium strength) far more rarely, actually only with paræsthesias and disturbances in sensation, therefore more symptomatically.

As local application, we consider also *counter-irritation* along the spinal column, particularly painting it with tincture of iodine, mercurial plaster (combined possibly with belladonna); also from the actual cautery (small point-size thermo-cauterizations on and near the spinal column, 20 to 30 at every sitting) one occasionally sees results, especially for the girdle feeling, etc. Furthermore, I wish to mention here the various methods of *stretching* the spinal cord, or the spinal column; either in the form of *suspension* (it is best to fasten to the seated patient a sort of Sayres suspensory apparatus at nape, chin, possibly also with shoulder straps, which by means of a weighted rope passing over a pulley, is drawn upward) or as so-called *bloodless stretching of the nerves*, by which—in various ways—the knees or the extended limbs of the patient, who is lying on his back, are passively brought as near as possible to the upper part of the body and kept thus fixed for some time. The hypotonia, which is usually present, facilitates this method, which, on the other hand, just because of this is not quite devoid of risk. The stretching procedures are praised very highly by a large number of writers. The so-called bloody nerve-stretching has been justly abandoned.

Massage is especially valuable for bed-ridden tabetics; it is particularly indispensable in the ataxic stage, partly to keep the musculature in good condition, and at the same time to prepare it for the exercise treatment, to be mentioned below, and partly as an aid to metabolic processes as a whole. To watch over the metabolism and the *state of nutrition* of the patient, is on the whole a main factor in the therapy of tabes. By every means, moderate forced feeding, nourishing foods, methods of stimulating the appetite, etc., the physician must try to keep the body in such condition, that it is capable of action and resistance; indubitably, thus alone, greater service is rendered the individual, but especially the more advanced tabetic, than by the use of complicated medicinal compounds.

The allotment of *rest and exercise* is of great importance. Above all, the slightest over-exertion on the part of the patient must be avoided. Accepting *Edinger's* "using up" (exhaustion) hypothesis, it would be a good thing to force the patients to take absolute rest for some time; *Edinger* himself has recently emphasized this again. But without considering the fact, that in the very protracted course of most cases, such treatment would be tantamount to an almost unendurable test of patience on the part of the sufferers, according to the views of most authors, the result of the rest cure is uncertain, even

in those cases, in which local over-exertion had certainly helped to bring about the disease (isolated ataxia of the right arm in tabetic clerks).

On the other hand, among the general therapeutic methods, *hydro- and balneotherapy*, rank next in importance after nourishment.

Almost all methods of procedure may be employed, especially in the earlier stages of the disease; they may be used for a long time, but, on the other hand, it should be a principle never to employ them in too energetic a form—especially in respect to the temperature. Few things can injure a tabetic more than using applications too extreme in cold or heat; I, myself, have seen several tabetics, who, after energetic schematically applied *Kneipp's* cures, showed a decided turn for the worse. Among the water "cures" that can be used at home, I recommend mostly the hip baths (as a rule, without shower) of 90° to 95° F., which are applicable in nearly all stages of the disease; alternating with them, or as a separate "cure" one can give artificial carbonic acid baths, pine-needle baths, to strong patients cold friction baths, to the weaker ones, lukewarm douches.

Not only upon psychic grounds, but especially because of their almost specific effectiveness, "bath cures" (watering places) are to be recommended, among which must be named first, the baths of Nauheim and Oeynhausen (carbonic acid thermo-baths); here too, however, baths of *medium* intensity are most effective. All other baths are inferior to these (but we might recommend in addition the sulphur baths—Aix la Chapelle, Neundorf—and the indifferent hot baths—Baden-Baden, Gastein, Wildbad); many tabetics, however, are much improved by simple *fresh air treatment* in the middle and high altitudes, whereas, sea baths, as a rule, are less beneficial.

The great variety and all too frequent ineffectuality of our therapy, is shown in the treatment of the separate tabetic *symptoms*, which we shall briefly discuss.

The *lancinating pains* are especially difficult to combat; as the attacks are usually short, the use of morphine is, as a rule, to be dispensed with, more so since these pains generally react best to the simple analgesic "anti-rheumatic" agents: aspirin, pyramidon, citrophen, lactophenin, especially antipyrin and antifebrin in the form of Erb's "mixed powder" with codein (antipyrin grs. 7, antifebrin grs. 5, codein gr. 1/3–gr. 1/4; instead of antipyrin possibly also phenacetin), migrainin, etc. If these agents are ordered in large doses, or in short intervals, the result is usually favorable. It is more difficult to combat the permanent pains, enteralgias, girdle-pains, etc., and most difficult of all the genuine tabetic crises, especially the *gastric crises*, which can be influenced almost only by large subcutaneous doses of morphine. The internal medicines above mentioned (and in addition, bismuth, anæsthesin, cerium oxalate, bromides, etc.), are usually ineffectual; I have occasionally seen results from lumbar puncture and from the injection of soda nitrate subcutaneously. If one does not

wish to give morphine, there remains, usually, only one way, namely, to let the crises spend their strength and to attempt to keep the patient, by complete rest and artificial nourishment, on a high plane of strength. Nor are the external applications of much avail here, whereas in lancinating pains, hydropathic packs, painting with iodine, chloroform, oil, methylsalicylate, salves and balsams may occasionally prove useful.[1]

The other sensory disturbances rarely become so painful or (except when trophic changes arise, cf. below), so dangerous, that they need a special therapy; labile faradic treatment, farado-massage may be used for par- and anæsthesias.

The *motor disturbances* with the exception of ataxia, likewise need no particular treatment; peripheral paralyses are rare; for the paralyses of the muscles of the eye, which incline towards spontaneous improvement, galvanization may be tried.

On the other hand, as is well known, the *treatment* of *ataxia* forms one of the main problems of modern tabes therapy, in the form of what is generally called the *Fraenkel "exercise therapy,"* which was variously tested and completed by *Fraenkel, Goldscheider, Foerster,* and others. As has been elucidated above, we understand by *ataxia,* speaking generally, and for practical therapeutic purposes, a disturbance of co-ordination, of the co-operation of the individual muscles and groups of muscles. To this corresponds the single therapeutic purpose: restoration of the lost co-ordination by *practising* this very process. Inasmuch as the disturbance evinces itself, according to our experience, preponderantly in the dark, that is, betrays itself in movements not controlled by the eye, the process, in practice, is very simple; the movement that has been disturbed is gradually learned again (by practice) under the control of the eye, until it eventually succeeds, even without observation. In our experience, the help of the eyes is a mighty factor in the movements of ataxic limbs, and allows, even without special practice, many movements to be performed, that would be wholly impossible, if the eyes were closed.

The exercise therapy is to affect every muscular area stricken by ataxia; as a rule, of course, first and most important, the legs. Since it must be a main principle, *that the patient is not to be tired out* (exhaustion may lead to outspoken deterioration in the condition of the patient), the first leg practice should take place in bed; from the most simple, there should be gradual advance to the difficult. For exercise therapy there exists a long line of useful directions (*Fraenkel, Goldscheider and others*) which can be used in complicated cases, especially also, to make greater variety possible. They describe also a great number of assisting apparatus (diagrams to be followed out by hands or feet, parallel bars to walk between, etc.), which, however, in the majority of cases can be dispensed with, especially if the physician, as

[1] Whether the results obtained by internal use of adrenal preparations, reported by *Röhmer* to the society of German neurologists, October, 1908, are confirmed, has not yet been ascertained.

should always be the case, watches over the execution of the exercises. As a rule every physician will be able himself to compose easily the most important movements. Besides the gradual learning of all movements, attention must be paid above all to the exact co-working of the single components of the most essential and important movements: walking, dressing, undressing, eating, writing, etc. Employing the necessary patience, the method leads to improvement in nearly every case, sometimes to an almost total disappearance of the ataxia. Of course, one must work with greatest consideration and care for weeks, often for months, in the beginning several times daily, for a few minutes. They are really *contraindicated* only during the first weeks of an acute, rapidly appearing and increasing ataxia and if the patient's general state of health is in too low a condition. During other intensive treatments (energetic bath cures) it is better to cease the exercise therapy temporarily.

The other symptomatic indications for treatment are more or less self-evident. A warning must be sounded against medication, directed to influencing tabetic *impotence;* all the highly recommended remedies, especially yohimbin, the effects of which are to be understood as a stimulation of the sexual sphere, have proved to be unnecessary, and partly injurious; in lighter cases, rest, a sparing therapy, together with general tonic procedures, is most likely to produce an improvement, which, to tell the truth, is usually transient. We can not give sufficiently strong warning against all extreme measures.

The *bladder disturbances* react relatively best to galvanization, occasionally also (bladder weakness) to ergot and strychnine. In pronounced retention, catheterization is, alas, unavoidable; this, after longer or shorter time, as a rule, causes an infection of the bladder, which, it is true, can be treated for some time by washing out, etc., but is apt finally to lead to ascending processes—pyelitis, pyelonephritis. In tabetic *retentio alvi,* medicines are to be used only in cases of extreme necessity.

The *trophic disturbances* are treated according to general surgical rules; the arthropathies can, as a rule, be treated only orthopædically.

Because of the usual, slowly progressive course of tabes, a *somewhat typical plan of treatment* may be outlined for many cases. To a large extent, however, this plan depends upon the stage in which the disease is recognized. If the recognition takes place, as it always should, at the beginning of the lancinating pains, for which the majority of tabetics consult their physicians, there should be introduced, under the above (cf. under *therapy*) conditions, a mercurial treatment, best by inunction. Naturally, besides this, at least during the recognizable advance of the disease, every active mental and physical exertion, especially exciting occupations, sports, hot or cold baths, sexual indulgence, should be immediately given up, or, at least curtailed. Mild hydrotherapy (hip baths) and galvanization can even now, but in any case, as continuation of the mercurial treatment lasting possibly four weeks,

be used for 2 to 3 months. In cases where the mercurial treatment is excluded (cf. above) an iodide therapy (iodide of potassium, iodipin, saiodin) is mostly to be recommended as a substitute for 2 to 3 months; it may be used also along with the Hg. treatment, or as its continuation. In the second place, after the course of the first 2 to 3 months, a generally tonic method may be employed in all cases: arsenic, cacodyl, *Erb's* tonic pills, etc., for 4 to 8 weeks. If circumstances permit, a "bath cure" should be interpolated in the summer, at Nauheim or Oeynhausen, with temporary galvanization.

Then in the normal course, a pause in treatment of some weeks or even months is to be recommended. After the symptoms have come to a standstill, one may wait to the end of the first year, and then carry out a similar program in the following years, with slight modifications (relative to hydrotherapy, the use of tonics, derivation to the skin, suspension). If the disease makes a marked advance, the therapy must naturally keep step with it, but with avoidance of every, even therapeutic exertion, for the patient; often absolute rest is now best for the patient. Symptomatic measures (especially exercise for the ataxia) naturally are to be employed at the same time. In the last stage of the disease, there is a question only of care and palliative measures, not infrequently of extensive morphine therapy. Even now, however, mild procedures, as galvanization, may be tried with some success. In any case, so long as no serious cachectic complications (serious pyelitis, bed sores) are present, there is need to beware as well of therapeutic nihilism as of meddlesomness, since in spite of the slight influence we are able to exert on the course of the malady, there are few diseases, in which such excessive demands may be made upon therapeutic resources as in tabes dorsalis.

(b) Friedreich's Disease (Hereditary Spinal Ataxia)

BY

S. SCHOENBORN (Heidelberg)

Friedreich's disease represents only a subdivision, though the best known and sharpest defined, of a larger group of *more or less systematized diseases of the central nervous system, which are of a degenerative nature, usually show hereditary or familial propagation, and are founded in part at least upon an anomaly of predisposition of the central nervous system, and develop from this.* Common to them all, is a more or less well developed degree of *ataxia*, which has given the group its name. To this are added, according to the individual form, disturbances of reflexes, scoliosis, bulbar symptoms, optic nerve changes, and disturbances in intellect. The best method of dividing the entire group may be that recently proposed by

Raymond, which divides it into 4 separate types of disease, of which we must recognize 2 as indubitably correct.

1. **Spinal form** or **Friedreich's** disease sensu strictiori, with ataxia, nystagmus, speech disturbances, absence of tendon reflexes, scoliosis, club foot.

2. **Cerebellar form** or hérédoataxie cérébelleuse (*Marie*) with ataxia, optic nerve changes, paralysis of the muscles of the eye, exaggeration of the reflexes, vertigo, intellectual disturbances.

Between these, there exist doubtless transitional forms as well as the combination of 1 and 2, which *Raymond* classifies as an independent fourth group: "type généralisé" of familial character. Whether an *independent* bulbar form (with dyspnœa, vomiting, cardiac arythmia) exists, seems to me doubtful from the literature; I myself have seen no such case. Raymond defines such a form as the last subdivision. Let us now turn to *Friedreich's* disease, in its stricter sense.

Friedreich's disease is of rare occurrence. In 1901 I could collect from the literature only a few more than 200 cases; meanwhile the number may at the most have doubled. Apparently the disease occurs disproportionately less frequently in Germany than in England and North America. Of its origin we know but little. It seems certain that it does not owe its development to a definite poison, differing in this from the otherwise so similar tabes, but that many factors may serve as "agents provocateurs." Very often the disease is familial, but it is very seldom directly hereditary. This may be due to the lack of possibility of transmitting the disease by heredity, since it is already far advanced, when the patients arrive at marriageable age, partly, as *Bouché* has it, the disease itself may not be transferable, but only the neuropathic tendency and degeneration, which (because of alcoholism, tuberculosis, syphilis) is actually found in the parents of the patients. It is probable that a slowly progressing inhibition in development of the spinal cord, which quite early is found very small, is the primary cause, which gives rise to the disease picture; the disease may then be excited by infectious diseases, possibly also by traumatism.

The disease begins very early, usually between the 6th and 14th years; all cases developing *after* the 25th year are doubtful.

The **pathologic-anatomical** conditions are rather typical. The spinal cord is noticeably attenuated, especially in the posterior portions (occasionally, also, in toto). To this corresponds an ascending degeneration of the posterior columns, always of the columns of *Goll*, occasionally partly, of the columns of *Burdach* also. As a rule, the lateral columns are affected also and most frequently the lateral cerebellar tracts, somewhat less frequently, the lateral pyramidal columns. The columns of *Clarke* and of *Gower* may be affected too, rather exceptionally the anterior pyramidal columns also. By French authors (*Déjérine*) emphasis is laid on the fact

that the posterior roots and the peripheral nerves degenerate as well (motor and sensory), which, however, is described as inconstant and insignificant by German authors (*Oppenheim*). The medulla oblongata and the cerebellum, however, do not show any changes in typical cases. The degeneration affects axis cylinders as well as medullary sheaths; in severe cases the axis cylinders in the columns of Goll in the cervical part of the cord disappear entirely.

The similarity to the process in tabes dorsalis, is quite considerable, if one regards, as do the French authors, the degeneration of the posterior roots as a regular thing, and the difference concerns particularly the lateral columns, which remain intact in tabes. But it is far more difficult to discover the pathogenesis of the disease. It is as yet quite doubtful whether the posterior columns are affected primarily, and everything else secondarily, or if the disease actually ascends from the periphery through the posterior roots. The latter could be accepted only with the aid of the hypothesis (*Déjérine*), that Friedreich's disease consists in a deficient development of certain neurons, together with insufficient replacement of the fibres used up in the course of time. This is contradicted, however, by the regular, intactness of the ganglion cells in the anterior horns and of the spinal ganglia, whereas in the columns of *Clarke* the ganglion cells are often found destroyed. Also the *Scherbak* hypothesis of primary degeneration exclusively of fibres passing to or from the cerebellum, is still lacking in pathologic-anatomical proof, however significant it might be in regard to the above mentioned principle of dividing this group of diseases. The pathogenesis of the individual symptoms we shall have to discuss later. On the whole, then, we shall have to consider Friedreich's disease in the pathologic-anatomical sense as a combined system disease with preponderant participation of the posterior columns.

The clinical picture of the symptoms generally corresponds to this. Nearly always the children themselves, or members of the family first notice the appearance of the *ataxia*. Their gait becomes uncertain, with legs spread wide apart; they stamp while walking; they spill the liquids they are carrying, and begin to write illegibly. If the movements are tested individually, a decided ataxia is seen to exist in the intentional as well as the unintentional movements, and it is usually equally pronounced in *all* the extremities at an early date; as a rule, the head also is attacked in the form of the so-called static ataxia: the patients sway even while at rest, i. e., in sitting or standing, rock constantly to and fro with the head and trunk, like a tree blown about by the wind. This instability affects all movements simultaneously, and reminds one of the ataxia of tabetics. But doubtless there is an admixture of another component (especially perceptible in walking), which reminds one of cerebellar unsteadiness. Very many patients exhibit *besides* the ataxia of the single movements, a wholly unsteady gait, as if intoxicated;

when walking they shove themselves forward, while they attempt, to a certain extent, to steer the body, which is usually held stiff. Occasionally there is even a certain degree of propulsion. With eyes shut, the unsteadiness, as a rule, increases, as in tabes, but the patients virtually never fall down. On the whole, because of the extremely slow development of the disease, the patients learn to control their movements in part, in spite of the ataxia. Thus one of my patients, in spite of a high degree of ataxia in the hands, was

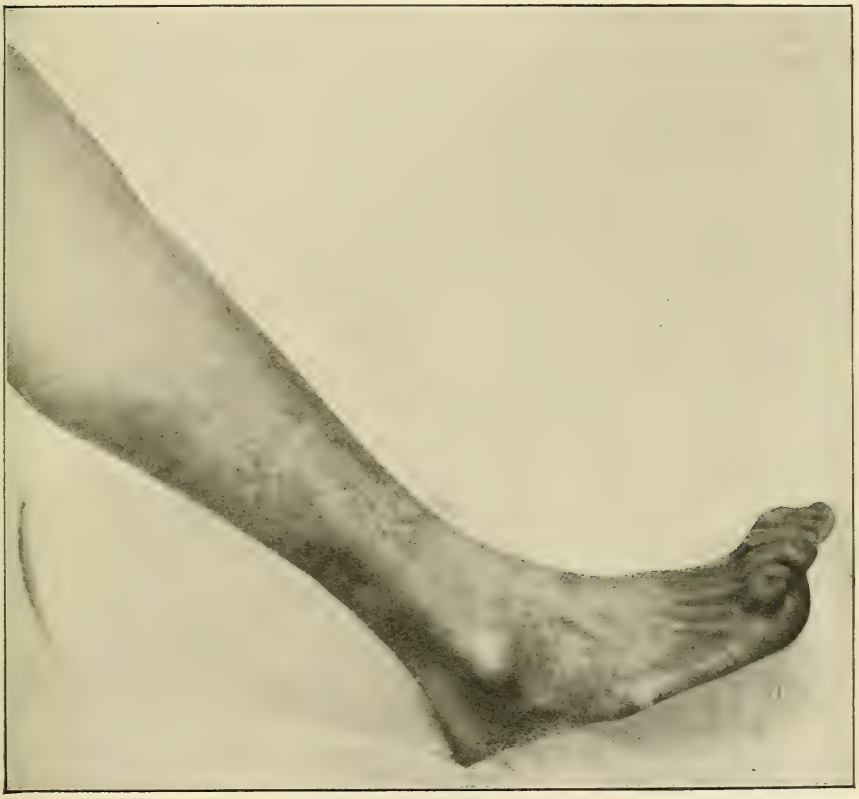


FIG. 61.—Form of foot in Friedreich's disease.

able to draw very beautifully and write very legibly, both, of course, very slowly.

Beside this disturbance in movement, there appears frequently, especially in the muscles of the head and face, also of the hands, a *muscular restlessness*, which recalls chorea (grimaces) or perhaps more often athetosis.

At the same time the mere physical strength of the patient is not decreased to any extent, though usually the arms and legs grow somewhat emaciated. But local atrophies are very rare, though the frequent pes varus (varo-equinus) seems to suggest them in Friedreich's disease. As a matter of fact, the planta pedis is usually very thin and the prominence of the arch of the

foot and of the tendon of the extensor hallucis longus makes the disease picture very peculiar, but I never found actual atrophy in it (which, however, is claimed for it by other authors), but rather an increase in the size of the extensor hallucis longus. There appears, moreover, corresponding with the prominence of this tendon and with the dorsal flexion of the great toe, an occasional phenomenon in the hand ("main bote") in the form of a hyperextension of the last phalanges. In this condition, slight muscular hypertension may be seen to exist in the parts affected, which otherwise is, however, usually absent in this disease. But, on the other hand, a considerable hypotonia of the muscles is scarcely ever found. To explain these otherwise astonishing, local muscular hypertensions, one must assume that they are the consequence of continuous cramp-like attempts at equilibration, in which the great toe plays the preponderant part. (The explanation of *Cestan*, that the *Babinski* reflex, usually present in the disease (cf. below) has, so to say, become permanent, is not sound) (Fig. 61).

Of other deformities we must mention the extraordinarily frequent kyphosis, or kyphoscoliosis, which is probably only a secondary phenomenon resulting from the lack of muscular balance.

Cutaneous sensibility as well as deep sensibility, are, strange to say, retained as a rule. Only isolated instances of disturbances have been reported in the literature; thus one of my patients made the characteristic statement, that when swimming, he did not know where his legs were; as a matter of fact, it turned out in the examination that coarser disturbances of the sense of position were not perceivable. *Oppenheim* mentions a similar case. Usually, however, both objective and subjective disturbances of sensation are wholly lacking. *Pains* also are scarcely ever complained of. The *tendon reflexes are absent* in all definitely marked cases. Of the cutaneous reflexes, the abdominal and cremaster reflexes are, as a rule, active, and the plantar reflexes show the form of the so-called *Babinski* reflex, slow dorsal flexion especially of the great toe, upon irritation of the planta pedis.

In regard to the cerebral nerves, one need only say that usually *nystagmus* is found, corresponding to intention ataxia, and that *speech* becomes uncertain early, now scanning, now explosive and above all it is distinctly slower than normal. The other cerebral nerves are normal in pure cases, especially those of the muscles of the eyes and the optic nerve. Disturbances of the sympathetic nervous system also (vasomotor disturbances, sphincter affections, crises) are in contradistinction to tabes, as rare as pronounced trophic changes.

The psyche remains normal. In one of my cases epileptiform convulsions were noted. As to combinations with bulbar symptoms, cf. below. Concerning the *pathogenesis of the individual symptoms*, it is to be remarked, that *ataxia* in its spinal components, is to be traced back to the process in the posterior columns; in its cerebellar components, to the degeneration of the

lateral cerebellar tracts. Whether the nystagmus is to be assigned to a cerebellar tract, remains doubtful; for the speech disturbance also, a clear anatomical foundation is lacking. The *Babinski* reflex we must probably ascribe to the (slight!) lesion of the pyramidal tracts. It is very striking, however, that notwithstanding the serious lesions of the posterior columns, no disturbances of sensation are present. It seems, therefore, that the columns of *Goll* are not absolutely necessary for the localization of peripheral sensations, and can even be replaced for their transmission; perhaps there is some connection here with the fact that in spinal ataxia the short fibres are retained, whereas the long fibres, contained for the most part in the columns of *Goll* are largely destroyed (*Déjérine*).

The **course** of the disease is very chronic, and it may drag on for 20 to 40 years. The symptoms increase slowly, but none of them in itself is apt to bring on death, which is usually caused only by an intercurrent disease.

Correspondingly, the *prognosis* quoad vitam is not unfavorable, quoad sanationem is absolutely bad.

The **diagnosis** of pure cases is easy. It is in itself easily distinguished from all pure system diseases; from tabes, additionally, by the lack of all disturbances in sensation and the absence of eye symptoms. Somewhat more difficult may be its separation from multiple sclerosis, the intention tremor of which may resemble ataxia; nystagmus, speech disturbance, youth, and often but slight spastic phenomena occur in it also. But, as a rule, here too, the difficulties are solved by the exact recognition of ataxia.

As great rarities some combined system diseases have been described, which, like Friedreich's disease, presented a combination of symptoms of the posterior and lateral columns, and in their course, much resembled it; but, as a rule, the spastic phenomena predominated in them, and the ataxia was slighter. Naturally, a sharp division is often impossible in these cases.

On the other hand, we have still to consider its demarcation from the disease picture of *hérédoataxie cérébelleuse* (of *Pierre Marie*).

In this disease, an ataxia develops in middle age, which exhibits an obvious but not always an exclusive cerebellar character. To this are added paralyses of the muscles of the eye, and nearly always atrophy of the optic nerve, frequently psychic disturbances (imbecility), exaggerated tendon reflexes. On the other hand, we miss the results of defects in muscular balance, which correspond more nearly to spinal ataxia—namely, talipes cavus, and scoliosis. The autopsy disclosed in pure cases of this disease an abnormally small cerebellum, occasionally also some diminution in the entire central nervous system, degeneration of the spinal tracts which stand in connection with the cerebellum and of the posterior columns. But these pure cases (also described by *Marie von Londe, Miura, Switalski, and others*) are still more rare than pure cases of hereditary spinal ataxia. On the other hand, in recent years, observations have been accumulating, in which symp-

toms of both diseases are combined: *Friedreich's* disease with psychic symptoms, optic atrophy, *Marie's* disease with spinal ataxia, kyphoscoliosis, talipes cavus. Even in one and the same family Friedreich's disease has been observed in pure and in mixed form (*Raymond*). Still rarer are the symptoms, which (cf. above) may be classified under the special group of the "bulbar form"; disturbances in respiration, in so far as they do not come from ataxia of the respiratory muscles, usually in the form of dyspnœa; cardiac disturbances, recalling myocarditis (which according to French authors, *Lannois* and *Porot*, frequently occur in the pure Friedreich as well), vomiting. That these symptoms occur when the disease process is continued to the medulla oblongata, is indubitably true; the formulation, however, of a "hérédoataxie bulbaire," seems nevertheless artificial.

Therapy, unfortunately, offers little, but luckily, with one exception, there is but little pressing need of it in Friedreich's disease. This exception concerns the removal of the ataxia, which, if it can not be entirely cured, can at least be vigorously combated by practice of the most important muscular movements in the form of *Fraenkel's* exercises. Very many patients, because of the gradual development of the disease, succeed by themselves, in extensively exercising the groups of muscles most necessary to their life. (For more details on *Fraenkel's* exercises, cf. under the article "tabes".) After all, therapy, except for a certain prophylaxis especially in infectious diseases in families of a neuropathic diathesis or in those already affected by Friedreich's disease, can proceed only symptomatically. In longer treatment, the same rules hold good as for the "tonic procedure" in tabes.

(c) Spastic Spinal Paralysis.[1] (Primary Lateral Sclerosis)

BY

FR. JAMIN (Erlangen)

The disease picture known as spastic spinal paralysis, the clinical expression of a purely motor system disease, a primary degeneration of the pyramidal tracts, is but rarely observed. It seems, however, that functionally the pyramidal tracts are very easily affected, like the posterior columns of the spinal cord, so that corresponding to their functional or anatomic injury, the symptom complex of spastic spinal paralysis, or of spastic paraparesis, temporarily or preponderantly steps into the foreground in many localized or diffuse diseased conditions of the spinal cord. But even though it seemed for a long time as if, according to anatomic findings, the sharply drawn clinical picture of the disease described by *Erb* in 1875, following up *Charcot's* hint, could not be characterized as a pure system disease, nevertheless in more recent times a series of clinically as well as anatomically thoroughly investi-

[1] Infantile spastic paraparesis (Little's and related diseases) is treated in the chapter entitled "Organic nervous diseases of childhood" by J. Ibrahim.

gated cases have proven that there is actually a primary motor system disease of the spinal cord, confined to the pyramidal tracts, with the symptoms of spastic paralysis, and thereby not only a symptomatological, but also a nosological individual place is secured for spastic spinal paralysis.

Symptomatology.—The spastic spinal paralysis of adults is, in its purely systemic form, a very protracted disease (extending for years and decades after the onset of the first phenomena), which usually occurs in the 2nd to 4th decade of life, progressing very gradually, but most constantly, and characterized by but few subjective disturbances. As a rule, the first phenomena appear in the lower extremities. The patients notice, when walking, that they become tired easily, and that a certain heaviness and stiffness in the legs interferes with the more active movements, especially such as running and jumping. Should their vocation make greater demands upon them, unpleasant sensations of weariness appear, occasionally, too, a feeling of cold in the legs (they are more than ordinarily difficult to move), but violent pains or paræsthesias do not occur. As the disease advances, the patients become more and more helpless; they can move forward only by means of small, dragging steps, and become so stiff in the legs, that when they meet obstacles, they are no longer able to avoid them quickly, therefore they stumble easily, and, occasionally, fall. A number of the patients can, with the aid of canes, preserve for an extraordinary length of time a limited walking ability; others, on the other hand, because of the increasing stiffness and weakness of the legs, and the supervening contractures, finally become bedridden.

FIG. 62.—Spastic spinal paralysis; posture in sitting. (*After Schoenborn and Krieger.*)

Examination enables us to recognize quite early the *spastic-paretic symptom complex.*

Particularly striking is the *stiffness* of the legs, the *hypertonia* of the muscles, which is exhibited even in conditions of rest, but more in active and more especially in passive attempts at movement. It leads to a functional stiffening of both legs, in the joints of the hip, knee, and foot, which occasionally is more marked at the beginning, on *one* side, than on the other. The stiffness prevails in the extensors, the gluteal muscles, the adductors, the power-

ful mass of the quadriceps muscles, and in the musculature of the calves of the legs. In advanced cases, therefore, the patients can not flex their legs sufficiently at the hips and the knees, to be able to sit down (Fig. 62). When trying to sit, the patient leans against the edge of the chair, almost as stiff as a stick, while the legs, almost at full extension at the knee, remain floating in the air. It is only with great effort and with the use of considerable strength, that passive flexion in the hip- or knee-joint may be carried out. Quickly executed attempts at passive movements merely increase the muscular resistance. In the ankle-joint, the stiffness may progress to such an extent, that, as a result of the "muscular joint ankylosis" (*v. Strümpell*), active and passive movements of the foot become virtually impossible. Frequently the extension of the leg as a whole is so forcefully produced, so fixed by muscular tension, that the patient can be lifted from a supine position, by *one* leg, almost like a board. Even in passive lateral movements of *one* leg, the pelvis and the other leg follow, whereas with careful yet forceful overcoming of the muscular tension, it may be demonstrated that the *possibility* of movement is retained in the apparatus of the joints and ligaments.

The fact that the patient, in spite of such an increase of the functional obstacles, is still able to walk for considerable distances in an upright position, shows that *no* decided *paralysis* of the legs is present. As a matter of fact, in movements of resistance, the muscles of the leg, and especially the extensors, prove, very frequently, to be still powerful, and we must ascribe preponderantly to the increased muscular tension, the fact that the active movements are so troublesome, so hampered and slow. Nevertheless, in many cases, a *diminution of the actual strength* of the legs is also demonstrable, independent of the obstacle of tension, especially in the abductors and flexors in the hip-joint, the flexors in the thigh (biceps, semimembranosus and semitendinosus) and in the dorsal flexors of the foot. This paresis of the flexors, i. e., the shorteners of the leg, combined with actually greater strength in the extensors or lengtheners of the leg, and the hypertonia prevailing in the latter, influences the *posture* and the *gait* in spastic paraparesis characteristically: the patients in order not to fall backwards, because of the extension of the hip, move the weight of the body far forwards, the outstretched hands at the same time seeking new points of support with the aid of canes or by grasping furniture (Fig. 63). Both legs remain in full extension, the feet in plantar flexion; and frequently the patients step only on the balls of the toes, until by standing longer, the weight of the body overcomes the tension of the muscles of the calf of the leg. The thighs are adducted, the feet turned slightly inward. In walking, the legs can scarcely be raised or flexed at the knee; in almost full extension, one after the other is pulled forward laboriously in a short circle, the tips of the toes are allowed to drag, or trailed forward and thus a short step is taken. At the same time, because of the overbalance of the adductors, the knees and the balls of

the toes frequently rub against each other on the medial side. Because of this the shoes are worn off, especially in the foremost part of the sole, and on the inner side of the tips of the toes. On sandy ground, slightly curved, almost parallel lines show the tracks of the short, dragging steps. And since walking is accomplished rather by dragging the extended leg along, the upper part of the body being bent forward, than by means of an elastic lifting swing, and replacement of the leg, as is performed by healthy individuals, it produces a characteristic rhythmic scraping noise in short intervals.

Next to the phenomena of hypertonia and paresis in certain muscle groups, the tendency to *associated movements* is characteristic of the spastic changes. Their common foundation lies probably in the fact that the movements of the legs, interfered with by weakness and increased muscular tension, are synergetically possible only by the co-operation of larger groups of muscles, trained to perform primitive movement complexes, whereas the volitional suppression of these combinations of movements, in favor of finer, isolated movements, which were learned during the course of life, though possible to the normal man, is now impossible. Especially clear does this disturbance appear in spastic individuals in the movements of the foot and toes. Whereas, in many cases, an isolated voluntary movement of the great toe, or of the entire foot, e. g., dorsal flexion can no longer be performed by the patient, strong active flexion of the limb at the hip or the knee is seen to be accompanied by a strong, insuppressible dorsal flexion of the foot, with conspicuous protrusion of the tendon of the tibialis anticus (*Strümpell's tibial phenomenon*). In lifting the whole leg, often also during attemps at walking, a very marked dorsal flexion of the great toe appears (*Strümpell's toe phenomenon*), which can not be produced isolated to the same excessive degree by active effort. Flexion of the leg at the hip, which is almost impossible when the leg is in extension, is also far more easily accomplished against resistance if at the same time the leg proper be flexed and the foot put in dorsal flexion, and, on the other hand, extension of the hip is difficult without simultaneous extension of the knee and plantar flexion of the foot.

The permanent hypertension and excessive excitability in individual muscles or groups of muscles, not only during voluntary functional activity but also to reflex stimuli, leads usually to a slight change in the permanent position of the lower extremities, to *contractures*. Usually the extended position of hip and knee is accompanied by plantar flexion with slight inward rotation and supination of the foot and dorsal flexion of the great toe, with moderate adduction of the thigh. In other cases, there appear, especially in long persisting affections, flexion contractures in the hip- and knee-joints in spite of the continuation of the equino-varus position of the feet and the prominence of the tendon of the long extensor of the great toe.

Without exception spastic spinal paralysis is accompanied by a marked

exaggeration of the tendon and periosteal reflexes in the lower extremities. Occasionally it is difficult to demonstrate this exaggeration of the reflex because of the enormous muscle tension, but even then it is usually possible with the limbs in a suitable position of rest; the patellar and a chilles tendon reflexes may be elicited; marked patellar and ankle clonus is very frequently

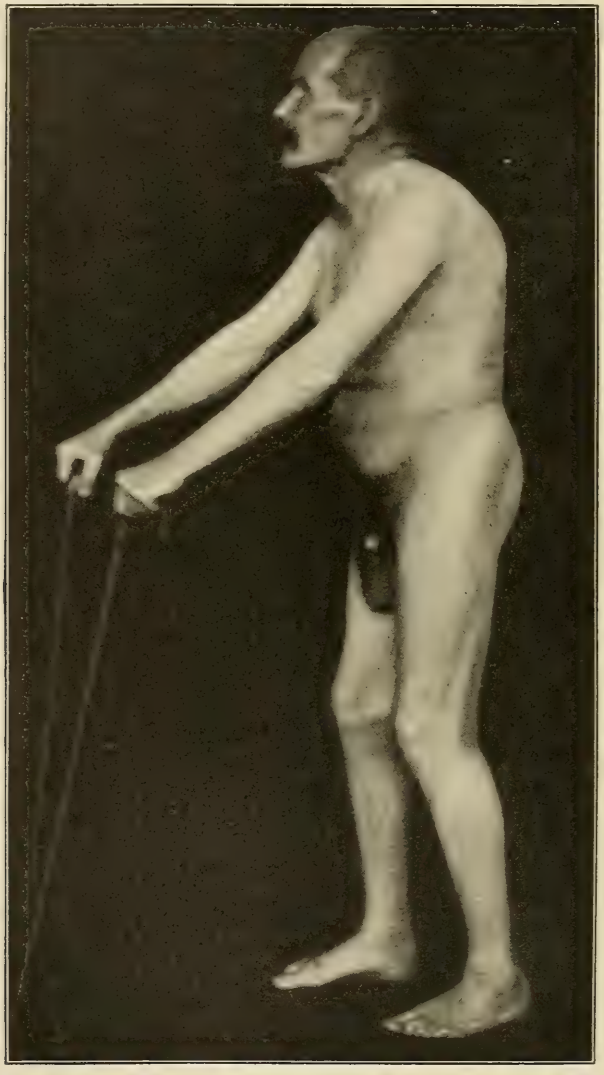


FIG. 63.—Spastic spinal paralysis; posture in standing. (*Erlangen Medical Clinic.*)

also induced in this condition. The reflexogenous zones are considerably enlarged; the reflex movements may occur on the other side as well, and in muscles at some distance. Not rarely, under the influence of weight alone, when the legs are hanging down, or after trifling spontaneous and reflex

movements, one leg, or even both, are thrown for some little time into clonic tremulous reflex movements, which, like genuine reflex spasms, can be suppressed only with considerable trouble by fixation of the limb for some time, or by vigorous stimulation of the skin. These spasms become very annoying to the patient. In standing and walking, also, there appears often, as a consequence of the abnormally vigorous reflex excitability of the muscles of the calf of the leg, and the peculiar position of the toes, a reflexly conditioned quivering and trembling over the entire body. Upon tapping the side of the dorsum of the foot, there frequently appears, instead of the normal dorsal flexion, plantar flexion of the toes (*K. Mendel*).

Of the *cutaneous reflexes*, those of the abdomen and the cremaster-reflex occasionally, appear to be diminished, but usually suffer no change worth mentioning. The cutaneous reflexes in the legs, especially those of the *sole of the foot*, are, as a rule, exaggerated and changed in a typical manner: lively flexing movements of the entire limb, or manifold combined reflex muscular contractions follow upon energetic stimulations of the skin. Especially characteristic is the *Babinski* sign, slow dorsal flexion of the great toe, upon irritation of the sole of the foot. This important evidence of spastic disturbance after lesion of the pyramidal tracts has, however, been absent in some cases of pure lateral sclerosis, or found in a less clearly defined form. The appearance of a powerful reflex dorsal flexion of the foot and toes upon irritation of the inner surface of the leg proper is also of great diagnostic importance (*Oppenheim's* dorsal leg phenomenon). Occasionally also, especially in younger patients, there appears, upon irritation of the sole of the foot, a spreading of the toes with particularly energetic abduction of the small toe (*Babinski's* fan phenomenon), which may, moreover, be combined with peculiar clonic twitchings of the toes.

In the *ascending progress* of the disease, the phenomena of hypertonia, paresis, and exaggeration of the tendon and periosteal reflexes extend also to the *upper extremities*. Then the arms, too, show changes in posture, with abduction of the upper arm, flexion of the forearm and pronation of the hand, while the muscular weakness is evinced preponderantly in lifting and extending the arm and on dorsal flexion of the hand. The arm reflexes are extraordinarily active; hand clonus may be brought about by brusque dorsal flexion of the hand and flexion of the fingers by tapping the back of the hand. The very rare skin reflexes in the arms, shown by abduction and extension of the arm, and spreading of the extended fingers upon irritation of the palm of the hand, have been but little studied.

Finally, the motor *cerebral nerves* may also be attacked; we may conclude that they are spastically disturbed from the presence of a strangely tense expression in the face, from difficulty in and inhibition of speech, from difficulty in the movements of the muscles concerned with chewing and swallowing, as well as from attacks of spasm of the glottis. In such cases, the loss

of central inhibition of expressive and emotional movements, is evinced in remarkable fashion by the appearance of *compulsory laughing and crying*, induced by slight stimulation and sometimes lasting without cessation, as long as one occupies one's self with the patient. When the disturbance of the entire central motor system of the pyramidal tracts is extensive, the disease usually shows a certain tendency to extend to the spinal motor centers. This may be recognized by the localized muscular atrophy that appears in places, especially in the hands (cf. also Fig. 64 *a*). These cases represent transitions to amyotrophic lateral sclerosis. In pure cases of primary lateral sclerosis, the conservation of the muscles and the absence of all localized atrophy is characteristic.

In *all* cases of spastic spinal paralysis, the functions of superficial and of the deep *sensibility* remain absolutely intact. *No vasomotor* nor *trophic* disturbances are observed, the organs of sense suffer no sort of injury, disturbances of the bladder and intestinal tract are absent. At most, when the spasms are very severe, possibly because of the participation in the general rigidity of the muscles of the floor of the pelvis, there may result difficulty in micturition.

Pathological Anatomy.—The symptom complex of spastic spinal paralysis is produced by a degeneration of the pyramidal tracts, which, in the pure cases, may be traced through the entire spinal cord, in the lateral columns, and even in the anterior columns (Fig. 64). The manner and duration of the disease determine the spread of this degeneration to the higher sections of the central motor-conducting tracts. Occasionally it extends beyond the pyramidal crossing to the pons, the cerebral peduncles and the internal capsule. The ganglion cells of the psychomotor fields of the cerebrum, are, as a rule, found intact, nor does the lesion extend to the motor ganglion cells in the spinal centers of the anterior columns, that is, their peripheral branches in the anterior roots and the peripheral nerves. Even the muscles themselves prove upon anatomic examination to be intact; in fact the muscle fibres are especially voluminous and hypertrophic in the regions most affected by the hypertonia and the exaggeration of the reflexes. Transitions to combined column diseases of the spinal cord may be observed, due to the presence of a slight systemic degeneration in the posterior columns, especially affecting the funiculi graciles (Goll), but in these cases, owing to the precedence in time and the permanent preponderance of the functional disturbances of the pyramidal tracts, the spastic paretic symptom complex comes so completely to the fore, that the phenomena of sensory loss of function, ataxic disturbances, and diminution in the tendon reflexes seem to be wholly concealed.

Ætiology.—The *familial* and *hereditary* occurrence of a pure spastic spinal paralysis, i. e., primary lateral sclerosis, observed by *v. Strümpell* and others, places the possibility of a definitely *endogenous* occurrence of this

systemic disease beyond doubt. But also in the isolated cases, probably a deficient development of the tract, which later is destroyed, gives the foundation for the elective disease, which is frequently excited in more mature age under *exogenous* influences, like intoxications, infections, nutritional disturbances (anemia), exhausting over-exertions, and traumatisms with their

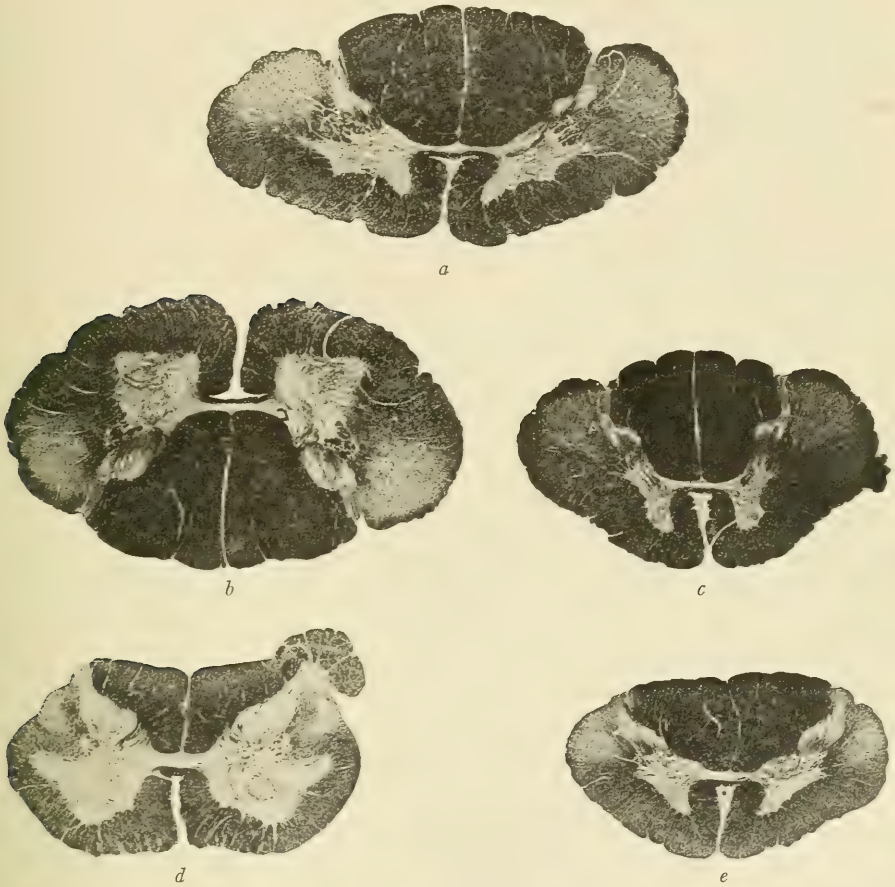


FIG. 64.—Primary pyramidal-lateral column sclerosis in spastic spinal paralysis. Weigert-Pal staining. *a*, Upper cervical cord. Besides the degeneration of the pyramidal lateral tracts a slight atrophy of the anterior columns exists; *b*, lower cervical cord; *c*, upper dorsal cord; *d*, lower dorsal cord; *e*, lumbar cord. (*Erlangen Medical Clinic.*)

consequent phenomena. The injuries produced as the after-effects of syphilitic infection, not infrequently remain for a long time confined to the pyramidal tracts; this is likewise reported of chronic lead poisoning, of eating lathyrus seeds (lathyrus sativus, lathyrus cicera, chick pea), of pellagra occurring in those living upon Indian corn, of phenomena consequent to the puerperium and to other infectious diseases.

Differential Diagnosis.—The clinical picture of spastic spinal paralysis is so characteristic in its combination of muscular rigidity, reflex exaggera-

tion, and typically distributed pareses and contractures, that it can scarcely fail to be recognized symptomatologically. The disturbances that take place in active and passive motility, the increase in the tendon reflexes and the pseudoclonus which are met with in hysteric and psychopathic patients (medico-legal accident patients) can not easily be mistaken for it if the arrangement and localization of the disturbances in movement are carefully studied, the cutaneous reflexes (*Babinski's* and *Oppenheim's* signs) carefully tested and the associated movements taken into consideration. In this respect serious difficulties in diagnostic judgment arise only when the organic disturbances are concealed or blurred by a hysteria that has supervened upon the organic disease.

More difficult and often answerable only after long and thorough observation is the question, whether the spastic-paretic symptom complex is produced by a *primary* pure lateral sclerosis, or whether we are dealing with a *symptom* of some other cerebral or spinal disease. The diagnosis of a primary lateral sclerosis can and ought to be made only, then, if after careful consideration of the subjective disturbances, and after minute examination of the sensibility, co-ordination, reflexes, the trophic and vasomotor relations, the spinal column and the cerebro-spinal fluid, the cerebral nerves and the organs of special sense (the ophthalmoscopic findings), the urogenital and intestinal functions, especially, too, after complete clearing up of the previous history, especially of the course of the disease, a symptomatic occurrence of spastic paresis in myelitis, compression myelitis, cerebro-spinal syphilis, hydrocephalus, spinal gliosis, progressive paralysis, *multiple sclerosis*, encephalitis and other diseases, may be excluded.

In *old age* there develops occasionally a spastic paraparesis with contractures, typical reflex changes and a disturbance in walking, which causes the gait to resemble that of the spastic because of its stiffness and weakness, which is accompanied by an attenuation of the pyramidal tract areas in the lateral columns and is probably brought about by nutritional disturbance in the spinal cord induced by arterio-sclerotic changes, though it may also have its seat in the cerebral cortex.

The **prognosis** of spastic spinal paralysis naturally depends upon the causes of the symptom complex. In the cases of pure lateral sclerosis, it is not unfavorable, in that while the course of the disease is progressive, it is frequently very protracted. The disease sometimes, too, may come to a standstill for years and, as a rule, does not affect the vital nervous functions.

The **therapy** can, to a certain degree, at least, be causal, inasmuch as it seeks to *prevent*, by rest and nerve economy, a further *exhaustion* of the weakened motor system. Therefore the patient, even at the beginning of the disease, must avoid all movements of the body, that stir him up to overhaste or over-exertion, or which by the tendency to make him fall, might

cause traumatic injuries. He must also avoid long walks and mechanical occupations requiring the frequent repeated use of certain muscles. On the other hand, by careful *practice* in walking and standing, at first, with the assistance of passive movement by stimulating the cutaneous reflexes, which often materially aid the flexing movements of the legs, the faculty for active movement can be improved methodically and by becoming accustomed to the altered circumstances, some degree of freedom in movement may be attained. It is desirable as well that the patient should become aware of the possibility of *substitution movements*, which may almost always be used, and perfect these gradually by use; for this, however, if he remains a long time in bed, he will lose both the opportunity and energy. Since, with the weakening of the cutaneous irritability and the lessening of the effect of weight in the warm bath, the stiffness in the muscles and the spasticity gradually give way, the persistent use of *protracted warm baths* is to be recommended. At the same time, the patients may derive benefit from the vivifying influence of carbonic acid baths at home, or at suitable health resorts. Careful *massage*, and especially passive aid in attempts at active movement, even in the bath itself, may further the motility, at least in the first stages of the disease. With the lowering of the tonic excitability by *medicines* (morphine, narcotizing injections into the dural sac) one will have to be most conservative, considering the duration of the disease. It is far more important to develop by suitable means the *active* powers of the patient in overcoming muscular resistance. For this purpose one may use moderate application of the constant current, while the stimulation of faradization and great changes in current are to be avoided. Spastic-paretic patients doubtless often recover their ability to move to a signal extent, if they leave externally unfavorable environments and go to a place where they can have good physical care and better nourishment.

The results of **orthopædic surgery** have not been, so far, very encouraging in spastic pareses, and especially in spastic spinal paralysis, unless we are concerned about getting rid of troublesome phenomena of secondary importance, as contractures. Neither through apparatus for support and posture, nor by cutting tendons and muscles, nor by plastic and grafting operations, can one effectively combat the disproportion between the increased reflex muscle tonus in some and the diminution of the voluntary development of strength in other groups of muscles. Recently, it has been attempted successfully, after laminectomy, to resect part of the posterior roots in corresponding selection, and thereby, without causing phenomena of serious functional loss, to weaken the reflex exaggerations and the hypertonia to such an extent, that the active capacity for movement becomes more free (Förster). In desperate cases, this method must be considered as the ultima ratio, if there be a possibility of strict aseptic procedure and most careful after-treatment.

(d) Amyotrophic Lateral Sclerosis

BY

FR. JAMIN (Erlangen)

The relatively rare disease, amyotrophic lateral sclerosis, affecting men more frequently than women, is a comparatively rapidly advancing destruction of almost the entire motor central nervous system, occurring in mature age—between the thirtieth and fiftieth years. In this affection not only the cortico-spinal motor conduction path perishes, especially in its most important component, the *pyramidal tracts*, but also the spinal motor centers in the *gray anterior horns of the spinal cord*, and the *nuclei of the motor cerebral nerves* situated in the medulla oblongata. In the clinical picture the injury of the first central motor neuron causes the *spastic paretic symptom complex*, as is shown in a pure form in spastic spinal paralysis, with muscular rigidity, exaggeration of reflexes and diminution in ability to perform voluntary movements—while the loss of the second peripheral neuron is responsible for the symptom complex of *paralysis* and *wasting of the muscles with the reaction of degeneration.*

One would think that the latter should bring about a disappearance of the spastic phenomena. But since to the extensive injury of the reflex inhibiting cortico-fugal motor paths, even in the advanced cases, there is opposed only a partial injury of the spinal and bulbar nuclei, which is confined to the innervation sphere and the trophic influence of special groups of muscles, even parts of muscles, there exists in amyotrophic lateral sclerosis an intimate *union of spastic symptoms with paralysis and so-called "degenerative" muscular atrophy*, which is the distinctive characteristic of this disease. Nor does the lesion affect all parts of the motor system in the same way from the beginning, but, on the contrary, according to the *localization* of the injury, definite *types* may be distinguished, which, in the majority of cases, denote successive *stages* of the disease:

1. The *paralysis* and *muscular atrophy* in the *arms* combined with *spastic* disturbances.
2. The disturbances in the movements of the *legs*, corresponding more particularly to the *spastic* symptom complex.
3. The *atrophic paralysis* in the area of the motor *cerebral nerves* appearing also with symptoms of *spasms, bulbar paralysis.*

Some observations of its occurrence as a *familial* affection, permit us to conclude that the **cause** of this exhaustion associated with degeneration of the motor nervous system at the time of active life, may rest upon a congenital weakness of this system. Physical over-exertions and the influence (interpreted with difficulty) of colds and traumatisms are stated as factors in exciting the disease. It is not known whether a specific uniform cause of the disease is predispositional to its development.

Symptomatology and Course of the Disease.—The insidious development of the disease makes itself in most instances first known by a functional disturbance of the upper extremities. In occasional cases, however, the disease, even at the beginning shows a *unilateral* type, and attacks the arm and leg on *one* side at approximately the same time. Even when it begins in the arms the disturbance on the *one* side (mostly the right) as a rule precedes that on the other by some months.

The patients themselves notice, that their arms and hands become weaker and less skilful at their usual work, that they are easily fatigued, without the occurrence, however, of pains or paræsthesias. In the further course, after some months, or only after a space of a year, there is noticeable a disturbance in gait, an unusual fatigue after walking, with stiffness, clumsiness, and weakness in the legs.

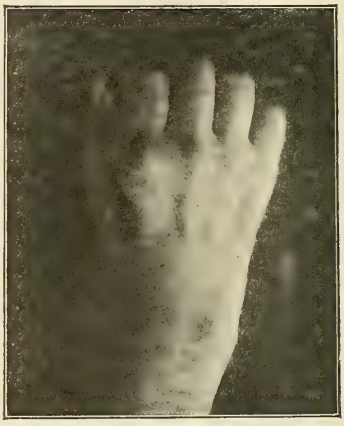


Fig. 65.—Hand in amyotrophic lateral sclerosis. Flexing contracture of the end phalanges, II–V. At the place of the adductor pollicis and the interossei visible hollows. (*Erlangen Medical Clinic.*)

The objective *findings* exhibit early in the arms *increased muscular tension* with a tendency to active contractures, especially with regard to an adduction of the upper arm, flexion of the forearm, pronation of the hand and flexion of the phalanges. Synchronously the characteristic *muscular atrophy* is found, at first, in the small muscles of the hand. The balls *of the thumbs* and of the *small fingers* become flatter. The wasting of the *interossei* gives rise to significant deep hollows on the back of the hand, the *adductor pollicis* becomes thin (cf. Fig. 65). Next, the *extensors in the forearm* become atrophic followed by the *long muscles of the thumb*, then, too, the rest of the muscles of the radial region and, since the flexors of the hand and the fingers are relatively better preserved and remain more effective, there results a *claw-like position* or flexion contracture of the fingers. Afterwards, the *deltoid* and the *triceps* become visibly atrophic, finally, the rest of the muscles of the arms and the shoulder girdle. Associated with the *muscular wasting*, there is a *muscular weakness* which ends in complete *paralysis*, and even the muscles which have retained their volume relatively well, frequently evince a considerable diminution in power.

Upon careful observation of the diseased muscular parts one sees, especially plainly in connection with attempts at movement, a lively play of *fibrillary* and fascicular *twitchings*. The *electric excitability* is lowered, but for the most part only in quantitative proportion to the wasting of the muscle fibres. In the more severely atrophied muscles, however, especially in

those of the hand, a *total* or at times only a *partial reaction of degeneration* may be observed.

The *tendon and periosteal reflexes* are always very *markedly exaggerated* in the weakened and atrophied arms. From the radius and the ulna, and from the tendons of the upper arm muscles, especially in the triceps, lively reflex muscular twitchings can be evoked; even wrist clonus is observed.

Sensibility remains absolutely *undisturbed* in all its qualities.

While in the arms, the muscular wasting and the paralysis preponderate early over the spastic phenomena, without wholly crowding them out, the *lower extremities*, should they share in the further course of the affection, exhibit the reverse. Here, the *spastic-paretic phenomena* step quite decidedly to the fore, whereas the muscular atrophy, fibrillary twitchings and changes in electrical excitability appear either not at all, or relatively late, and then mostly in the muscles of the leg proper. The laborious, dragging, sliding gait, the paresis of the flexors, group action of the extensors and flexors of the limb, and associated movements (tibial and toe phenomenon) spontaneous clonus and marked increase of all tendon reflexes, changes in the cutaneous reflexes (*Babinski's* great toe reflex and *Oppenheim's* leg reflex), finally, too, contractures unite with the general hypertonia of the muscles of the leg, which is made still greater by skin stimuli and motion, to make the clinical picture of *spastic paraparesis*. In the trunk, and the legs, also, disturbances of sensation are altogether absent. With the exception of a slight difficulty in urination and defecation (constipation) due to spasms in the musculature of the pelvic floor, there appear no bladder or intestinal disturbances.

After the patient, by a combination of spastic paraparesis and a more or less complete inability to use the upper extremities, has already reached a very helpless condition and has become bedridden, the disease becomes really tormenting and even dangerous, when, after *one* or *two* years, the *last group of symptoms*, which, in many cases, however, has developed much earlier, namely, *that of bulbar paralysis*, is added to the other symptoms.

Now, with the paralysis that appears in the area of the motor cerebral nerves, and the muscular atrophy, *speech disturbances* develop; speech becomes first nasal and difficult to understand, is articulated worse and worse, finally becomes *aphonic;* there appear *difficulty in swallowing, paralysis of the soft palate*, of the *tongue* with visible atrophy and fibrillary twitchings, of the *muscles of chewing*, and of the *facial muscles*, especially in the lower half of the face. The *lips* become thin and can no longer be raised and pursed at will, whereas the expressive movements of the face are frequently retained. In these regions also *side by side with* and often *before* the *appearance* of atrophy and paralysis, the *loss of cerebral inhibition* makes itself felt in many ways: the masseter reflexes are exaggerated; the muscles of the face are tense, often spasmodically distorted; *glottis spasms* appear, sudden and apparently causeless bursts of emotion (*obsessional laughing and crying*) with retention

of the intellect indicate the increase in reflex excitability. In the wrinkled tongue, which is moved with difficulty and in the lips and the muscles of the pharynx the *reaction of degeneration* or at least slow contraction upon galvanic stimulation may be demonstrated.

Only the muscles of the eye are, for the most part, spared. The mechanism of *respiration* may be impaired by the co-affection of the *diaphragm* and the *muscles of the ribs*, so that the patient finally succumbs to a respiratory paralysis, unless the secondary phenomena of paralysis of the throat and the swallowing apparatus (aspiration pneumonia) and the disturbances in nourishment, necessarily connected with the bulbar disturbances, have already caused death after a few years.

The course of the disease and the order of the appearance of the individual symptoms differ in different cases, depending upon whether the lesion of the pyramidal tract or the loss of the motor nuclei in the medulla oblongata and in the spinal cord has temporal precedence, and according as to which retains the preponderant importance for a time, or permanently. Thus, there are found *transitional forms to spastic spinal paralysis*, in which the atrophic paralysis remains altogether undeveloped, to *progressive muscular atrophy*, in which, from the beginning, the paralysis with muscular wasting conceals the spasticity, or stops it in the final stage of the disease, and to *progressive bulbar paralysis*, in which, early, the paralysis of the cerebral nerves, accompanied by slight spastic disturbances in the extremities, comes to the foreground.

Pathological Anatomy.—Transverse section of the spinal cord (Fig. 66) shows in amyotrophic lateral sclerosis very distinctly, especially in the cervical segment, the combination of *atrophy* of the *pyramidal tracts* with *atrophy* of the *anterior horns*, corresponding to the spastic atrophic paralysis which is most pronounced in the arms. The *pyramidal tracts* have wasted in the lateral column and, occasionally, also in the anterior column, not here only but in their entire spinal course, and in their ascending way through the medulla oblongata, the pons up to the cerebral peduncle and the inner capsule. Slight wasting of fibres, too, is noticeable in the rest of the area of the antero-lateral column, in some cases, also in the medial posterior column, without the appearance, however, of noticeable sensory phenomena of functional loss. The *gray matter of the anterior columns* becomes reduced, especially in the cervical segment, by the wasting of its network of nerve-fibres with the exception of the reflex collaterals (*Oppenheim*) and by destruction of a large part of the large motor ganglion cells. The anterior roots, too, usually become smaller. A corresponding *nuclear wasting* may be observed also in the bulbar centers of several *cerebral nerves*, in the nucleus of the *hypoglossus*, the *vagus-accessorius*, the motor part of the *trigeminus*, and in a slighter degree also in the *facial*. The psychomotor cortical fields and their nearest connection paths, also, have not always been

found intact. Slight proliferation of the supporting connective tissue replaces the nervous elements that are lost.

To the extensive loss of the spinal motor centers, correspond *degenerations in the peripheral motor nerves*, which, however, are difficult to find in the mixed nerve trunks, and also because of the mingling of fibres still capable of conduction with those that are already incapable. In the atrophied *muscles* of the arm, the *wasting of muscular fibres*, resulting in some places in complete loss of the contractile substance with increase of the muscle nuclei and the interstitial connective tissue, may be demonstrated

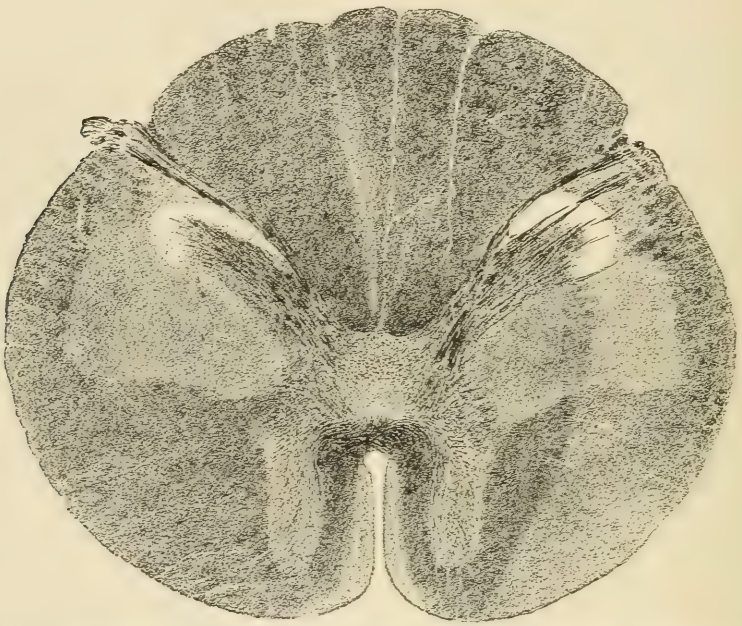


FIG. 66.—Amyotrophic lateral sclerosis. Weigert staining. Uppermost cervical cord. The pyramidal tracts show degeneration in the anterior and lateral columns. A slight degeneration is to be found also in Goll's column. The gray anterior columns have almost disappeared. (*After Schmaus.*)

easily, accompanied under certain circumstances by profuse infiltration of fat. In some muscles, affected only by partial atrophy, more or less atrophied muscular fibres are mingled with those that are well preserved.

In the **differential diagnosis** of amyotrophic lateral sclerosis it is especially important to note that this disease is strictly confined to motor functional disturbances. Testing the sensation and consideration of the subjective troubles, which are not to be explained by disturbances in movement, like pains and paræsthesias, make easily possible a distinction from the other spinal troubles, which like syringomyelia, spinal tumors, cervical pachymeningitis, myelitis localized in the cervical segment, or syphilis dna tuberculosis of the cord may be accompanied by similar atrophic paralyses

in the arms with spasticity in the legs. Differentiation from the rest of the system motor diseases is made possible by the subacute course and the *commingling of spastic phenomena with cervical and bulbar atrophic paralyses*. In regard to this the increase of the periosteal reflexes in the atrophied forearms is especially significant for the myatrophy connected with lateral sclerosis; it does not appear in like manner in spinal muscular atrophy, nor in other lesions restricted to the anterior horns, due to spinal gliosis or hæmatomyelia. In *multiple sclerosis*, which not infrequently simulates motor systemic diseases, localized muscular atrophies are scarcely to be observed in similar arrangement and development; it may be recognized frequently, besides, by proof of disturbances in co-ordination and due consideration of the eye findings even in the atypical cases.

The **prognosis** of amyotrophic lateral sclerosis is in every respect unfavorable. Irresistibly, and apparently indifferent to external influences, the disease proceeds to its fatal end.

The **treatment** can therefore only be symptomatic. Consideration and good care are obviously necessary because of the helpless condition of the patient. In the final stages, the patients, under certain conditions, have to be fed through a tube. The spasticity and contractures may be combated by *baths*, careful *massage*, also by suitable bandages; because of the progressive nature of the disturbances, it is better to abstain from operative treatment. The employment of the *constant current* in the region of the nape and along the spinal column, also in the area of the atrophied muscles, and the administering of nourishing foods and strengthening medicines, etc., is to be all the more recommended since only steadily continued therapeutic procedures may spare the patient, who with unimpaired intellect is gradually perishing, a clear insight into his condition.

(e) Spinal Progressive Muscular Atrophy (Duchenne-Aran)

BY

FR. JAMIN (Erlangen)

The *spinal motor centers in the gray anterior horns* of the spinal cord with their thick network of nerve fibres and the groups of motor ganglion cells are the last central terminal stations for the *muscles*, through which the stimulations emerging from the superior centers of the brain, the cerebellum and the subcortical ganglia, as well as the reflex impulses created in the spinal cord itself must take their way. Their destruction causes a secondary degeneration of the peripheral motor nerve fibres and a complete *exclusion* of the attendant contractile muscular fibres from *every voluntary and reflex activity*, that is, not only a cessation of the motor activity, which has visible results, but the loss of every function, and therefore also of the tension that

exists in times of rest. The consequence of this loss of all functional stimulations, is, for the muscle, a considerable diminution of the processes necessary to its existence, of circulation and nourishment. Since such a muscle now lacks the power of an up-building restitution to the natural loss of its substance, a power afforded it by its normal specific function, it perishes after the loss of its innervating and thereby trophically acting spinal center, in a slow way and, if considered for short periods of time, scarcely perceptibly, yet irresistibly, until the complete process of destruction, atrophy, is reached.

An *injury of the anterior columns* of the spinal cord with the corresponding secondary phenomena in the *muscles—paralysis, loss of reflex excitability* and *muscular wasting*—occurs in numerous diseases of the spinal cord. Even among the *system* motor diseases of the spinal cord it has already been mentioned as a partial phenomenon of amyotrophic lateral sclerosis. *Pure primary degeneration* of the spinal motor *centers*, the premature using-up and gradually progressing destruction of these important motor central stations is, however, observed in the very rare *disease picture of spinal progressive muscular atrophy*.

An entirely sharp separation of the manifold forms, under which a congenital weakness of the nervous and muscular apparatus of motion appears clinically, can certainly not be made. As we have seen that the injury of the cortico-fugal motor tracts often does not halt when reaching the spinal and bulbar nuclei, but changes these also functionally and morphologically, just so the atrophic processes of the myopathic forms of progressive muscular atrophies, which begin in the muscles, lead not infrequently to an impairment of the function and the anatomic condition of the gray anterior horns. Therefore, even in this respect, in a large number of *transitional forms*, which frequently make the interpretation of an individual case more difficult, the theory of the unity of the entire motor system, possibly also a unity of the pathological processes holds good. Notwithstanding this, a considerable number of observations during a protracted course of the disease and with characteristic grouping of symptoms, the peculiar localization of the muscular atrophy, made sure by anatomic findings, justifies the demarcation of *progressive spinal muscular atrophy* as a different group of diseases from the related affections, *amyotrophic lateral sclerosis* and *myopathic muscular atrophy*.

There is but little known about the *causes* of the disease. Its appearance in several members of *one family*, that is, upon a common *congenital* predisposition, has been observed. The disease occurs, however, more frequently isolated, in adults, and more frequently also in men than in women. The *exciting* influence of local or general *traumatism* is certainly of importance, but very difficult to judge because of the unusually slow and insidious development of the disease. This is likewise true of the influence of colds and in-

fectious diseases. It seems that the disease may develop upon the foundation of a *syphilitic infection*, just as in tabes the injury of the nervous system not infrequently extends itself to the spinal motor nuclei. Severe physical work and *over-exertion* certainly further the development of the disease, but one must beware of a confusion with the muscular atrophies in *occupational neuroses* (*Oppenheim*) which are more favorable, prognostically and much less severe. Finally, the development of a progressive spinal muscular atrophy in more mature years, has been likewise observed being grafted on a defect which developed in childhood as the result of an anterior poliomyelitis.

Symptomatology and Course.—It is especially characteristic of progressive spinal muscular atrophy, that it does not at one fell swoop or with giant strides seize upon large sections of the spinal motor centres, but beginning very gradually in a restricted area, proceeds to the disjunction of one central element after another, and therefore gives time for the development of the *trinity resulting from wasting of the ganglion cells*, of the *motor nerve fibres* and of the *muscular fibres*, before new and further areas are attacked. Therefore we see, that in most instances the phenomena of *paralysis* and *muscular atrophy* appear *simultaneously*, and that years often pass before a larger part of the muscles of the body participates in these disturbances.

FIG. 67.—Effeminate hand in spinal progressive muscular atrophy. Atrophy of the ball of the thumb, of the little finger, and of the lumbricales. (*Erlangen Medical Clinic.*)

Nearly always the symptoms of functional loss *appear* in the region of the *small muscles of the hands*. The patients themselves probably notice that they no longer possess their former skill in finer movements, and that they can no longer develop sufficient strength in grasping, reaching for, and holding objects. Upon examination at this time, one finds even then an emaciation and flattening of the *ball of the thumb* and of the *little finger*, often more markedly developed on the right than on the left hand. Especially the muscles of the *thumb*, first, the *opponens* and *abductor brevis*, then also the *flexor brevis* and the *adductor*, lose their functional ability and become thin, limp, and weak Early, the long muscles of the *thumb* in the forearm, the *extensor* and *abductor pollicis longus* are added, so that then the thumb begins to lie almost immovable on a plane with the metacarpus (*ape's hand*) (Fig. 67). But the other small muscles of the hand are not spared: the atrophy of the *interossei* and the *lumbricales* causes a prominence of the lines of the bones on the back of the hand and in the palm, and together with the

paralysis of the extensors of the terminal phalanges and the contracture of the still more effective long flexors and extensors in the forearm, leads to the form known as *claw hand* (main en griffe). In the period following, the *extensors* of the *forearm* are affected, also the flexors, the atrophy appearing preferably in the *ulnar division of the flexors*, so that here, too, the contours of the bones become more and more prominent. At the same time, there often exists and persists a peculiar diminution of the extremity towards the *distal* end, which is, moreover, favored by the wasting of the cushion of fat, which is mostly connected with the atrophy of the muscles (bird arms). Associated with the wasting of the muscles which continues for years, and the greater and greater immobility of the hands, the *bones*, too, become thinner, and appear in a skiagraph poor in calcium bone-substance, somewhat wasted, with extremely delicate structural design, without usually changing their general essential configuration.

The further spread of the atrophy and the paralysis does not ascend the arm *gradually*, but usually by leaps, as a rule, jumping next to the *deltoid*. With the emaciation, at first partial, then total, the patients lose more and more the ability to raise their arms (Fig. 68). The arm hangs down from the shoulder, adducted, the use of the hands is made possible with difficulty only by the assistance of the muscles of the upper arm—which may for a long time continue to act—and by swinging motions or by the combined employment of the forearms pressed against each other. Furthermore, the atrophy spreads into the region of the *muscles of the shoulder*, early in the *supra- and infra-spinati*, so that the contours of the bony shoulder girdle, especially of the scapula, become more and more prominent; then, too, in the *cucullaris*, whereby the raising of the arm and the turning of the shoulder are made still more difficult; in the *rhomboidei*, the *latissimus dorsi*, the *pectorales*, and the *muscles of the back of the neck*, so that the head sinks forward and can not be voluntarily raised. If the atrophy of the muscles is very far advanced, as a result of the functional loss of the muscular tonus, the ligaments of the joints relax, and the joints become loose. Finally, a participation of the *muscles of the trunk* may make the upright position difficult, and lead to a compensating lordosis.

The *lower extremities* are but rarely affected by the paralysis and the muscular wasting; but when the affection is of long standing, there appears, here too, and particularly in those *muscles* of the *leg* that raise the *foot*, and in the *pelvic muscles*, a flaccid atrophic paralysis. Therefore the gait becomes a waddle, due to the loss of fixation of the pelvis to the thigh. In order to lift the foot, which is hanging limply down, from the ground, the thigh must be raised abnormally high. The patients become fatigued easily and become more and more helpless. Even in spite of the advanced degree of atrophy, the patients can help themselves to a certain point and retain some mobility through the skillful use of the muscles

that are retained and still act; finally, however, such a large part of the musculature of the body is affected that the patients become bedridden, and depend upon the help of others for all that has to be done. *Participation in the disease process of the muscles of respiration* may finally lead to serious difficulty in breathing, and thereby to a fatal end. The condition becomes

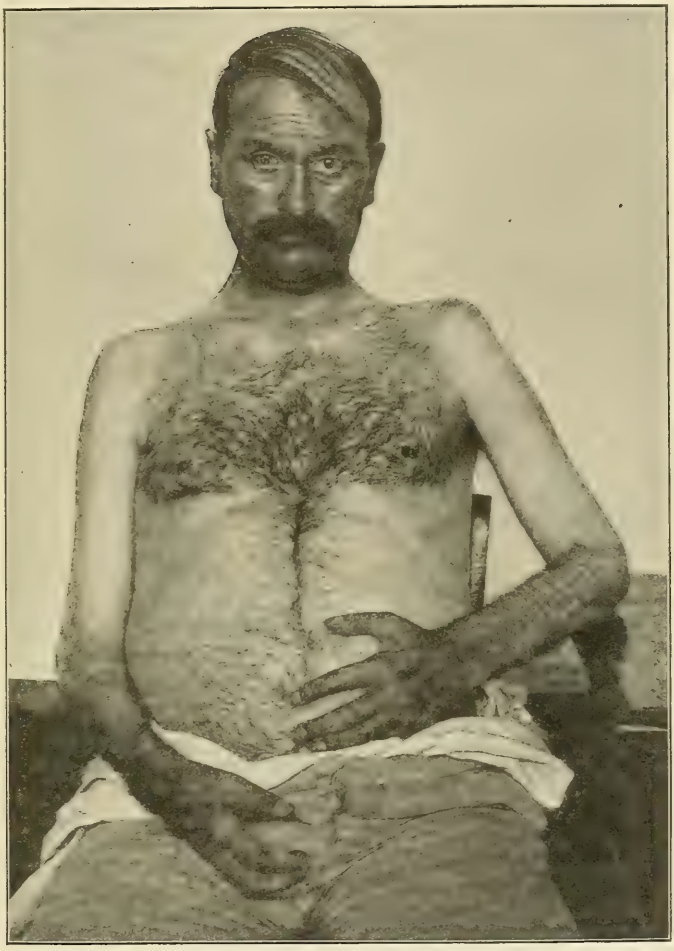


FIG. 68.—Spinal progressive muscular atrophy. Besides the little muscles of the hand the muscles of the forearms and the upper arms have also atrophied. (*After Schoenborn and Krieger.*)

equally dangerous when the muscular atrophy reaches the realm of the motor *cerebral nerves* and leads to the development of progressive *bulbar paralysis* (cf. p. 318).

All these stages of the disease develop in the course of years. The reacquisition of the capacity for movement, when it has once been lost, is impossible; an improvement is possible only in so far as the patients may learn

by practice and habit, in adaptation to the changed circumstances, to put the muscular parts that have been retained to better use. Quite often also, the condition remains apparently stationary for a long time. But if, at intervals of some time, an exact and detailed examination is made, one will usually be able to note an advance of the disease.

The onset of the disease in the arms, i. e., in the *small muscles of the hand*, and next to them in the *deltoid*, is of by far the *most frequent* occurrence. But cases are observed also, in which the first signs appear in the *shoulder girdle* or even in the *legs*, and they are then much more difficult to find, than the characteristic change in the configuration of the hands, which immediately attracts the notice of the skilled practitioner. Possibly in this selective action, so far as *localization* of the disturbance is concerned, the *kind of occupation* may play a certain rôle, so that the regions most employed fall first and most completely under the sway of the atrophic process.

The *paralysis* remains, as a rule, confined to the *atrophied* muscles, and, in its degree, is dependent upon the degree of muscular wasting which frequently spares isolated bundles of fibres in the individual muscles, as can occasionally be seen clearly in the contraction of the deltoid.

In addition to the *muscular wasting*, which is usually confined within strict boundaries and in typical localization, and the parallel *functional disturbance* of paresis, or paralysis in certain parts of the limbs, some further changes are worthy of notice.

The *tendon and periosteal reflexes* are absent in the *atrophied* regions. In the arms they are nearly always lost or diminished to a high degree, and this is the case even when the musculature in a part of the fore- and upper arm is relatively well retained and is functionally intact. In the lower extremities they are usually retained for a correspondingly longer time. The cutaneous reflexes are changed only in so far as muscles that have become wholly atrophied, naturally can no longer be stimulated to contract.

In the muscles attacked by the atrophic process, unless the atrophy is complete, *fibrillary and fasciculary twitchings* very frequently appear. These are occasionally so active in the small muscles of the hand at the beginning of the disease, that they may lead in the quietly resting hand, to small jerky movements of the fingers; for instance, of the thumb. They are especially striking in the deltoid, in which the muscular restlessness may be increased to a long and persistent wavering and oscillation of the individual fibres and bundles of fibres, which may be still further increased by stimulations of the skin, tapping the muscle, under the influence of cold and electric stimuli, as well as by active attempts at movement.

The *electrical excitability* of the wasted muscles, which only in rare instances exhibit a tendency to fatty infiltration, is *quantitatively* always lowered. In complete atrophy, well localized tests in which powerful currents are

employed and the often very disturbing influence of a communicating loop of the currents in the muscles that are intact is carefully avoided, may demonstrate the presence of *total reactions* of *degeneration*, especially in the small muscles of the hand. More frequently, however, the muscles, which are usually considerably reduced in size, present, corresponding to the merely partial atrophy, only a *partial reaction of degeneration* with *sluggish* contraction upon *direct* galvanic stimulation.

Sensibility, which should always be thoroughly tested, remains completely normal in all its qualities. The paralyzed arms and hands become cyanotic, as a consequence of hanging down immovably, and feel cold; the skin over them occasionally becomes thin and delicate, but decided vasomotor and trophic changes are never found. *Bladder disturbances*, *disturbances* of the *genital functions* and of ability to empty the bowels, do *not* belong to the picture of the disease.

The occurrence of *myotonic* phenomena is worthy of mention (muscular rigidity after active movements, increased mechanical excitability of the muscles and myotonic electrical reaction) in cases of disease which, at least, according to the localization of the simultaneously occurring muscular atrophies seem very similar to progressive spinal myatrophy. Probably, it is a matter, however, of atrophies, which proceed from the preceding myotonic change in the muscles and are not dependent upon a primary spinal localization of the process.

Pathological Anatomy.—Anatomical examination in progressive muscular atrophy reveals a *wasting of the gray anterior horns* in the spinal cord, confined mainly to the cervical segment. The horns are reduced in size; many ganglion cells and nerve fibres disappear. The ganglion cells still present are noticeably changed in structure. The *anterior roots* are atrophied and their medullated fibres considerably diminished in number. In pure cases the medullary sheaths in the spinal cord are *intact*. On the other hand, there is a loss of motor nerve fibres in the *muscle-nerves* that belong to the injured segments, and distinct changes of structure may be seen to exist in the *muscles* themselves. The muscle fibres are pale in muscles showing a high degree of atrophy and show, in microscopic sections, great narrowing and proliferation of the nuclei. A luxurious growth of connective tissue surrounds the thin muscle fibres, which up to the most extreme degree of atrophy, under otherwise normal conditions still permit us to recognize transverse stripes, but finally become reduced to very thin, slightly granular, clouded little fibres, which no longer show any obvious structure. Frequently fibres of considerably better preserved size and sharply defined structure, lie among the muscle fibres, which have wasted to a considerable extent or even so completely as to leave the sarcolemmic sheath almost empty. The *muscle spindles*, corresponding to their physiological import as organs of the *deep sensibility*, suffer, in the pure spinal muscular atrophy, no change.

A real "*degenerative*" change of the *muscle fibres* (fatty degeneration, wax-like changes, fissures and cracks in the muscle fibres) does not occur in spinal muscular atrophy, which, as contrasted with myopathic muscular atrophy shows only *gradual* anatomical differences. Nor has the occurrence of genuine *hypertrophy* of individual fibres within the muscle, which has become seriously atrophic in consequence of loss of innervation, been determined with any degree of certainty. The anatomic-histological examination of the muscles, and especially of little pieces of muscle, taken from the living body or from the dead body before or during rigor mortis is extremely complicated by the changes occurring in the surviving contractile substance during the preparation, and by the variable changes in the condition of the muscular protoplasm dependent on metabolism and toxic influences before death. The *functional* changes observed in the atrophied muscles (electrical reaction of degeneration) following disjunction of the spinal motor center are dependent upon the destruction or degeneration of the end branches of the peripheral *motor nerves, not* upon the *morphological condition of the muscle fibre.* The term "*degenerative muscular atrophy,*" which is useful, in contradistinction to muscular atrophy where nerve conduction is retained, is therefore not to be understood, in a pathologic-anatomical, but *purely* in a *functional* sense, or in relation to the condition of the nerves. It concerns an atrophy, which goes hand in hand with those changes of mechanical and electrical muscular excitability, which can be proved to exist in "muscles, the nerves of which have been destroyed."

The **diagnosis** of spinal progressive muscular atrophy is based upon the protracted *course of the disease*, extending over years and decades, upon the *parallelism* between *paralysis* and *muscular wasting*, upon the typical *localization* and the *leap-like advance* of the symptoms, the *absence* of the *tendon reflexes* and all spastic phenomena and the strict *confinement* of the disease picture to the *motor sphere.* Especially does consideration of pains and of sensory disturbances make it possible and, as a rule, quite early in the disease, to distinguish it from *spinal gliosis* or *cervical compression myelitis;* it is in respect to muscular changes alone that they may give rise to similar clinical pictures. We may refer here to what was said under amyotrophic lateral sclerosis. From this latter muscular atrophy is separated by the absence of spasticity in the lower extremities and of all reflex exaggeration. The distinction from typical cases of *myopathic dystrophy* is made possible in most cases by the localization in the small muscles of the hand, by the fibrillary twitchings, and by reactions of degeneration, but, especially in respect to localization, there are many transitional types. The course of the disease and the succession of the symptoms allow us to easily avoid confusion with *multiple neuritis*, and with the *acute poliomyelitis* of adults. *Chronic poliomyelitis*, which is closely related to spinal muscular atrophy, might furnish most of the differential-diagnostic difficulties, were it not for the fact that

domination of the paralyses over the atrophic defects, and a broader extension of the process over larger masses of muscles, from the very first speak clearly for poliomyelitis.

As an *infantile hereditary* and *familial form of progressive muscular atrophy*, cases of quickly progressing muscular atrophy have been reported *Werdnig*, *Hoffmann*) which appear in *earliest childhood* in *brothers and sisters*, attack the *muscles of the thigh*, of *the pelvic girdle* and *the trunk*, then, also, of the *extremities* and lead to deformity of the vertebral column. The *atrophied muscles* show *reactions of degeneration*. The children, in the first years of their life, yield to the progressive muscular wasting as a result of their congenital liability to intercurrent diseases. The anatomic findings show *degeneration of the cells of the anterior horns* and of the peripheral nerves.

The **prognosis** of spinal progressive muscular atrophy is always *unfavorable* to this extent, that recovery or a permanent checking of the disease is impossible; the illness always shows a tendency to spread, even though slowly. In this way the condition of the patients becomes really helpless, even if they do not often suffer for a long time directly from the disease. A dangerous turn through the appearance of bulbar disturbances, of paralysis of respiration, or of intercurrent diseases, favored in their effect by the difficulty of the patients to move, are, at all times to be feared.

Therapy is virtually powerless in face of the *progressive tendency* of the disease. One must be satisfied with avoiding a furthering of the progress of the disease, by reduction of bodily movements, and guarding against all over-exertion, to which the patients, precisely because of their weakness, are very easily exposed. Careful *treatment* by *exercises*, arranged to suit the *peculiarities* of each case, *massage* and the use of the *constant current* may serve to strengthen fibres still capable of functioning, in the muscles that have already been attacked and to awaken new efficiency in them. It is important to combat in this way a premature stiffening of the limbs by a secondary contracture of the antagonists (which have become too powerful) by *strychnine* (daily, subcutaneously up to gr. 1/60) or with injections in the paralyzed muscles. An attempt at *medicinal treatment* by strychnine preparations administered in other forms, possibly combined with iron or arsenic, may be recommended. If some suspicious factors speak for a previous case of *syphilis* (serum test, scars on the skin or on the mucous membranes, lymphocytosis of the cerebro-spinal fluid), a long and energetically conducted treatment with iodide of potassium, with or without careful mercurial treatment must be attempted, since, though it cannot restore the musculature that has been lost or cause it to regenerate, it checks, under certain conditions, further development of the disease, or of other nervous manifestations thereof.

(f) Neuritic Progressive Muscular Atrophy

Peroneal, Forearm Type of Progressive Muscular Atrophy (Charcot-Marie, Hoffmann)

BY

FR. JAMIN (Erlangen)

Among the many and various forms, in which systemic change of the nervous system may express itself preeminently by myoatrophic conditions, a group of diseases, not very frequently observed, may be set aside from the closely related pictures of spinal and pure myopathic atrophies, which usually, if not always, appear familially and in childhood are characterized by the localization of degenerative muscular atrophies in the distal parts of the body, in the foot and leg, the hand and the forearm, and by slight disturbances of sensation prove the participation of the sensory conducting paths; in most cases these changes predominate in the peripheral nerves.

This neuritic progressive muscular atrophy begins very gradually in later childhood, in some cases, however, not until adult life. It attacks, as a rule, several members of a family, sometimes through several generations, sparing neither boys nor girls, but showing a distinct preference for the former. Isolated cases occur as well. If the disease has attacked a family, it may skip individuals or entire generations, only to reappear among their descendants.

The disease, then, is decidedly endogenous, developing upon congenital foundations. Accidental ætiological factors—getting drenched, and catching cold, traumatism, post-infectious cachexias and intoxications—may be said to be only exciting causes of the disease, or to assist in its development.

The arrangement of the **symptoms,** in particular the localization of the myoatrophy, is very characteristic. The patients themselves become aware of their disease by the remarkable weariness and weakness of the feet in walking, and also by the change in the contour of the feet, which interferes with the use of shoes.

Paresis and circumscribed muscular atrophy, discovered in the examination, appear first in the *small muscles of the foot;* they cause the contours of the bones upon the dorsum to stand out, the sole is deepened, and the medial edge of the foot is flattened out. Early, too, appear paralysis and atrophy of the muscles of the leg that are supplied by the *nervus peroneus*, above all, of the *extensor hallucis longus*, then, too, of the *extensor digitorum* and of the *peronei*, also of the *tibialis anticus*. The anterior and external sides of the leg, therefore, appear emaciated and through this atrophy raising the foot, especially its outer edge, becomes impossible, likewise dorsal flexion of the toes, above all of the great toes. When the leg is raised, the tip of the foot, therefore, as in peroneus paralysis, drops towards the floor with the claw-like

toes in plantar flexion. In the beginning, and occasionally for a considerable period of time, the foot remains easily, even abnormally movable at the ankle-joint; it swings loosely in the ankle-joint in passive movements. But since, as time goes on, the contrast in efficiency of the atrophied extensors of the foot and toes and the considerably better conserved muscles on the flexor side of the leg, of the group of the calf muscles, becomes more and more pronounced, *contractures* form (cf. Fig. 69). There develops, symmetrically on both sides, a formation of pointed and club foot, that can no longer be made normal, a *pes equinus varus*. Since in the course of the disease, the triceps suræ is usually also (gastrocnemius and soleus) attacked by the atrophic process, so that the curve of the calf is lost, the plantar flexion of the foot may also be lost; the contracture position is then controlled by the long flexors of the toes and the *tibialis posticus*, which often continues powerful for a very long time; when the action of the peroneal muscles is completely lost, it causes the foot to turn inward with the formation of "hollow foot." Then the toes can no longer be passively extended. The patients now walk upon the outer edge of the foot, even upon the outer dorsal side of the foot, upon which callus forms, and since the patients are mostly young individuals, the tarsal bones may be crowded out and moved from their natural places.

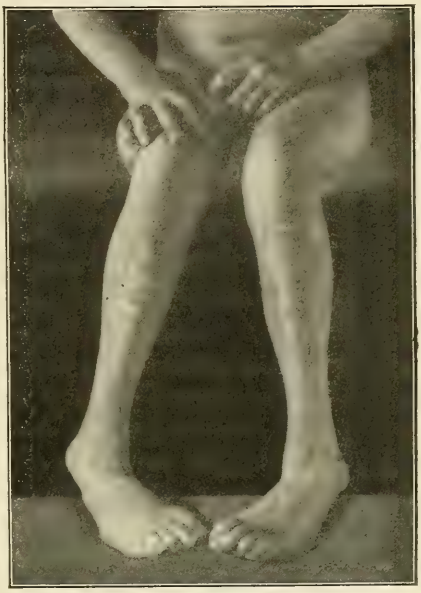


FIG. 69.—Posture of the feet in neuritic muscular atrophy. (*Erlangen Medical Clinic.*)

The muscles of the thigh become atrophic late and to a lesser degree. On the other hand, there appears earlier, in rare cases, even preceding the paralysis in the feet, an atrophy and corresponding paralysis of the *small muscles of the hand* and the *extensors of the forearm* with claw-like formation of the hands and tapering of the distal division of the arm, as it is described in spinal muscular atrophy. In later stages of the disease, the muscles of the upper arm and shoulder participate also, to a moderate degree, in the muscular wasting, whereas the muscles of the face and the trunk, also the bulbar realms are mostly spared, and therefore the muscular atrophy in these cases rarely reaches such proportions as to seriously threaten health and life.

The atrophied muscles show occasionally, but not regularly, *fibrillary twitchings*. Their electrical excitability is, both upon indirect and direct stimulation, lowered quantitatively, corresponding to the degree of the

atrophy; under certain conditions it is entirely lost. In the regions that are more severely affected, the peroneal muscles, the small muscles of the hand, etc., the presence of partial or total reactions of degeneration may be demonstrated.

The *tendon* and *periosteal reflexes* are, always, so far as the atrophy extends, frequently, too, throughout the extremities, wholly *lost* or considerably weakened. The cutaneous reflexes are not changed, in so far as they are not necessarily restricted by the loss of muscular regions that have become atrophic, or by the contractures.

Sensibility is really not much disturbed, but in many cases it does not remain perfectly normal; painful sensations and paræsthesias are observed in the atrophic limbs, which can not be explained by the changes of position and movement alone, also dulling of the superficial sensibility to the point of complete anæsthesia in the feet. The feet are cyanotic and feel cool. More serious trophic disturbances do not develop. Bladder and rectal disturbances are also absent.

The **pathologic-anatomic** investigations have shown that this disease is dependent upon degenerative changes in the peripheral nerves, which supply the atrophied muscles, especially in their distal intramuscular ramifications. Atrophies in the corresponding segments of the gray anterior columns of the spinal cord have also been found. Likewise the posterior columns (funiculi graciles Goll) have been found to be the seat of chronic changes. The changes in the atrophied muscles correspond to the picture outlined in spinal muscular atrophy—serious muscular wasting with connective tissue changes of the muscle parenchyma, while fatty infiltration is rarely present, and genuine muscular hypertrophy occurs but seldom in the well-preserved muscles, which in part are strengthened through their compensatory activity.

The **diagnosis** is based on the beginning of the disease in childhood, the very protracted course, the peculiar localization of the symptomatic muscular wasting, which resembles spinal muscular atrophy of the peroneal-forearm type. An absolute strict differentiation from spinal muscular atrophy can not always be made because of the anatomic changes which in many respects are identical, and there are transitions even to myopathic dystrophy, which, however, as a rule, begins from adjacent groups of muscles. Genuine polyneuritis develops far more quickly into a paralysis, which has occasionally similar localization, but is usually far more extended, displays more striking sensory disturbances (pains, ataxia) and, after a stormy course, permits the return of ability to move, while neuritic muscular atrophy, in its symmetrical, gradually progressing extension, produces irreparable defects. The continuous increase in the wasting of muscles, lasting for years, from slight beginning in the periphery, distinguishes it from poliomyelitis also.

A disease similar to this, with similarly localized muscular wasting, the same disturbances of electrical excitability, and slight sensory disturbances,

has been described under the name *interstitial hypertrophic progressive neuritis;* this disease may also be familial. It is distinguished from the peroneal form of muscular atrophy by the objectively demonstrable thickenings of the peripheral nerves (peroneus, ulnaris) as well as by the complication with distinct disturbances in co-ordination.

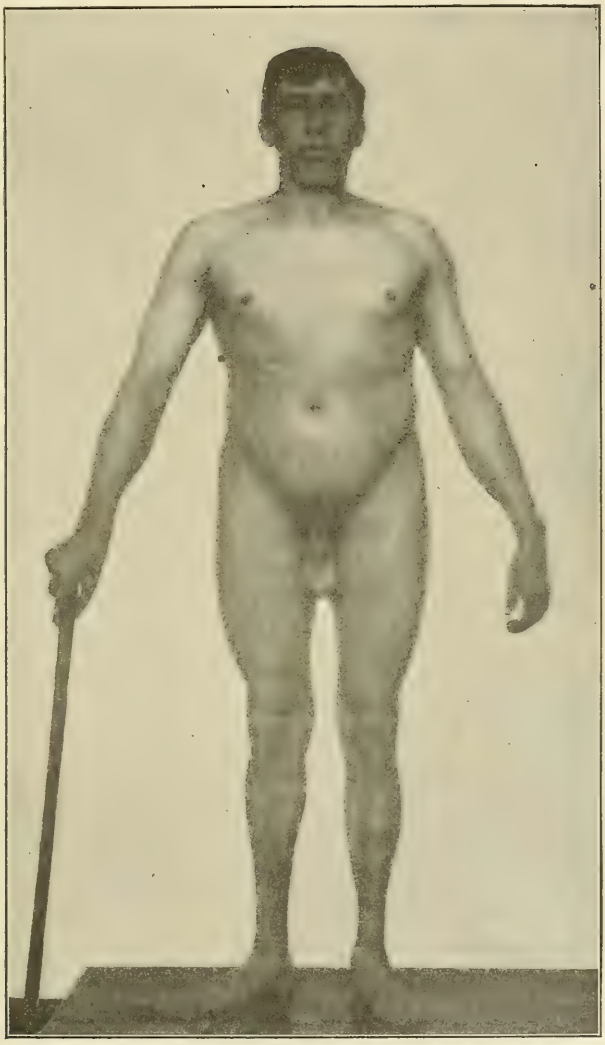


FIG. 70.—Neuritic muscular atrophy. Pronounced atrophy of the peripheral parts of the extremities, especially in the leg proper. (*After Schoenborn and Krieger.*)

The **prognosis** with regard to life is not really unfavorable. The ability to move and the possibility of following one's vocation, are, however, always to a great extent impaired, especially when the upper extremities also are severely attacked.

Therapy cannot stop the wasting of the muscles. By massage and

electrical treatment, as well as by a diligent care of the entire body, the rate of the progression may be somewhat influenced, and the condition of the patient is made distinctly more endurable by a careful strengthening of the retained muscles by exercise and use. The changes in form and posture of the feet, which are most disturbing, cannot be removed permanently or checked by supporting apparatus. The function of the preserved muscles, especially of the tibialis posticus, always forces the foot, if it is to be used at all, into an abnormal position again. As in all progressive muscular atrophies, plastic operations also afford little prospect of permanent results. The gait and posture of the patient may be best improved, by first loosening the tendons shortened by contracture, and then anchylosing the ankle-joint into a correct position permanently by an operation (arthrodesis), in order that the action of the better preserved muscles of the pelvis and the thigh may be given a safer support in walking.

(g) Subacute and Chronic Poliomyelitis

BY

FR. JAMIN (Erlangen)

In rare cases, in adults, a disease is observed, strictly confined to the peripheral motor system, associated with muscular wasting, which, in the localization of the paralysis and the muscular atrophy, as in the developing sequence of the phenomena, can be distinguished from spinal progressive muscular atrophy, as well as from the acute inflammatory diseases of the anterior columns and the peripheral motor nerves.

Probably subacute and chronic inflammatory processes are concerned, together with degeneration and atrophy in the gray anterior columns of the spinal cord, which owe their origin to the selective effect of *chronic poisonings*. Lead poisonings, autointoxications, diabetes mellitus are considered causal factors. It is possible that traumatic shocks favor the peculiar localization of the process of the disease. In the pathologic-anatomical picture, chronic, inflammatory changes in the vessels, as well as extensive degenerative-atrophic changes in the ganglion cells and in the mass of fibres of the anterior horns, have been demonstrated.

In the **clinical picture** the **symptoms of paralysis** are most prominent. Immediately, without any signs of a general disease, there usually appears first a weakness in the legs, which is increased in the course of a few days to a widespread paralysis of the extremities, but which, nevertheless, usually remains incomplete, attacking some groups of muscles more severely, and sparing others. The extent of the paralysis is not confined within definite bounds. In irregular succession one limb after another may be attacked, or both legs may be paralyzed alone, or both arms. After weeks, and months in some cases, all four extremities may have become paralytic, nor do the

muscles of the trunk remain intact; and in the decidedly progressive cases, there finally appears a paralysis of a bulbar character, which together with a paralysis of the muscles of respiration may lead to death.

Only after the serious motor functional disturbance has been present for some time, does the "*degenerative*" *muscular wasting* become apparent in the

FIG. 71.—Poliomyelitis anterior chronica. Severe atrophic paralysis of both hands; at the left side also atrophy of the deltoideus and the brachio-radialis. (*After Schoenborn and Krieger.*)

paralyzed regions. The muscles decrease in volume quickly and to a considerable degree, become limp and weak. As a rule they show *fibrillary twitchings*. The electric examination permits us to recognize a diminution of the electrical excitability and partial or total *reactions of degeneration*, even in paralyzed, but as yet slightly atrophied, muscles. The tendon reflexes are markedly

diminished or entirely abolished, and the cutaneous reflexes are lowered to a degree corresponding to the paralysis.

The sensibility in the pure cases, remains intact in all its qualities, should there be no more diffuse injury of the spinal cord. In particular, stronger sensory phenomena of irritation, also trophic disturbances, and disturbances in the urogenital and intestinal innervation are absent.

The disease does not always progress to paralysis of all four extremities, the trunk and the motor cerebral nerves. After weeks, it may become stationary, with more or less extensive paralytic and atrophic defects remaining non-retrogressive, or, after months, there may possibly be an extensive improvement of the condition, even recovery.

In regard to **differential diagnosis,** the common polyneuritis is of importance, since it has caused confusion with chronic poliomyelitis. But it is distinguished from this by the course, which is mostly more rapid, and above all, by the appearance of sensory phenomena of irritation and of loss of function, severe pains, sensitiveness of the limbs and the nerve trunks to pressure, and disturbances of sensibility and ataxia. In spinal muscular atrophy, and neuritic muscular atrophy, there develop the more sharply circumscribed functional disturbances, corresponding directly to the degree of the muscular wasting, and progressing simultaneously from fibre to fibre with it. They progress far more slowly than chronic poliomyelitis, which in quick succession, at once paralyzes entire groups of muscles.

The **prognosis** may be doubtful in the beginning of the paralysis. If the functional disturbance soon remains confined to a smaller region, and, after the development of the atrophy only partial reaction of degeneration sets in (middle form) the chances for recovery are not unfavorable. But if prolonged investigation shows that the flaccid atrophic paralysis marches on and on and can not be checked, it is to be feared that the vital muscular regions will be affected, and that the disease will be fatal.

The **treatment** in the advanced cases must be guided by the principles that hold good for acute poliomyelitis and spinal muscular atrophy. At the beginning of the paralysis, rest in bed and very little exercise of the muscles is suggested; massage too is to be applied only very cautiously; in any case galvanic treatment with the anode, whose action is not so energetic, is to be preferred to the use of faradic currents. Only when the phenomena retrogress noticeably, may the regeneration of the nerves and muscles be furthered by persistent methodical treatment with the constant current. Where there is some suspicion of a toxic ætiology, one should endeavor to eradicate it by dietetic measures, sweating and baths, also by medicines (iodide of potassium, preparation of salicylic acid) with exact control over the excretions. The possibility that similar forms of disease may develop after a syphilitic infection ought to be considered in the treatment.

(h) The Combined System Diseases of the Spinal Cord

BY

FR. JAMIN (Erlangen)

The detailed anatomic investigation of the endogenous, congenital, primary column diseases of the spinal cord, which are characterized by hereditary and familial occurrence (*Friedreich's ataxia*, spastic spinal paralysis) showed that not infrequently, a combined lesion of several systems in the lateral and posterior columns of the spinal cord proved to be present, even though the clinical picture of the disease was distinguished by the predominance of functional disturbance in *one* system group; in spastic paralysis, of the pyramidal tracts; in *Friedreich's ataxia*, of the posterior columns and the cerebellar lateral columns. It was natural, therefore, to assume that pictures of symptoms, which allow us to conjecture the presence of an approximately similar functional disturbance in the various sensory and motor tracts of the spinal cord, have also been caused by a primary selective injury in the fibre systems of the posterior *and* lateral columns, whether this injury be based upon a peculiar inborn weakness of this system, relative to its being used up easily, or to general noxæ, or whether it is caused by the specific effect of toxic influences on these systems of tracts. As a matter of fact, there are a series of observations, in which the clinical picture of the disease permits us to recognize almost exclusively a functional disturbance of the long tracts of the spinal cord, especially of the pyramidal anterior columns, the pyramidal lateral columns, the posterior columns, and the cerebellar lateral columns; and the anatomical findings also seem to establish the assumption of a pure primary combined columnar disease. But if the undoubtedly primary endogenous columnar diseases, which are discussed in the chapters on *Friedreich's ataxia*, and *spastic spinal paralysis*, are left out of consideration and also the many varied combinations of tabetic phenomena as being an exogenous disease, the pathogenetic relations for the isolated cases of clinically demonstrable combined columnar diseases, are found considerably more complicated.

Pathological Anatomy.—The more recent investigations by *Nonne* and *Fruend*, *Henneberg* and other authors, have shown that in most of the cases, the detectable degenerations in the spinal cord correspond, on the whole, to the course of the long fibre systems of the pyramidal tracts, the posterior columns, and the lateral cerebellar tracts, but that they are not strictly confined to these systems, but spare, in part, definite sections, especially those directly adjacent to the gray substance and partly extend irregularly at various levels of the spinal cord in various ways, and often asymmetrically, beyond the area of the well-known tracts. These variations in the spreading of the degeneration over the cross-section of the individual column systems, cannot be quite satisfactorily explained by the individual

differences in the development of the physiologically differing systems. In addition, transitional types, from apparently purely systemic columnar degeneration to decidedly focal disease of the spinal cord, occur.

In those places, in which the degeneration in the posterior and lateral columns is most marked, in the dorsal or cervical segments, dense sclerotic glia growths appear, in place of the degenerated posterior columns and in the center of the lost pyramidal lateral column, whereas the similarly destroyed lateral cerebellar tracts are replaced by a looser net-work of glia fibres. In the midst of the scleroses, however, thickenings, hyaline degenerations and perivascular infiltrations or scleroses are found on the *vessels*, which seem to confirm the supposition that it is not a matter of *primary degeneration of the columnar fibres*, but of the cicatricial *end-result of focal diseases of the white substance emanating from the vascular system.* By the confluence of small focal lesions in the white matter, there arises, associated with the phenomena of ascending and descending degeneration, from the picture of a disseminated myelitis the picture of a visible, but by no means strictly confined columnar disease. For the peculiar arrangement of the focal disease the white matter's scant supply of blood-vessels from the small branches of the marginal arteries of the spinal cord is the cause, in the area of which, naturally, with slighter disturbances in circulation, sclerosis of the fines tramifications and similar injuries appears more easily than in the far more richly supplied gray matter. Nevertheless here, too, as in other injuries which affect the cross-section of the spinal cord over a considerable space, the increased vulnerability of the long paths, especially of the posterior columns, and the pyramidal tracts may influence the arrangement of the degenerations, and especially the distributions of the functional disturbances. It is to be considered also in comparing the anatomical findings with the clinical picture of the disease, that the *anatomic* picture, especially in so far as it is confined to the degeneration of the medullated sheaths alone, and does not affect the make up of the axis cylinders, is not able to fully explain the *functional* disturbances. This explains also why under certain conditions, when there are extensive anatomic changes in the spinal cord, only slight functional disturbances are observed, and vice versa. The lateral cerebellar tracts only participate systemicly in the pathological process, if the degeneration of the lateral columns in the upper dorsal portion of the cord is considerable, because, in that case, the fibres passing from *Clarke's* columns to the lateral column are injured on their way through the focal injury in the lateral column. *Clarke's* columns, themselves, are often found intact; in other cases, they too have become atrophic, probably through retrograde degeneration.

If, therefore, it is probable, that the combined columnar diseases are caused, not by a primary selective injury of the tracts, but by disseminated focal disease as a consequence of disturbances in circulation, ischæmia, or

by an injury in the white matter, the result of a blood-vessel disturbance, yet in many cases, the preference for the posterior columns, the pyramidal tracts, and the lateral cerebellar tracts gives these processes of degeneration a peculiar impress, and justifies us in regarding these forms of disease, which differ even in the clinical course from the acute myelitic processes, as a separate group.

Ætiology.—The causes of the pathological changes described, which are rarely examined anatomically until the stage of the secondary and the resulting cicatricial conditions, are manifold. From a number of infectious and toxic injuries; from septic, chronic nephritic and tubercular processes, from pellagra, alcoholism, diabetes, carcinomatosis, lead poisoning, combined columnar diseases may result. Syphilis, too, may evoke a similar disease picture. With special frequency, extensive lesions seem to develop, in serious anæmic and cachectic conditions such as in pernicious anæmia, and leucæmia. In many of these constitutional diseases, special importance attaches probably to the disturbances of circulation, caused by arteriosclerotic changes in the smallest vessels, and it is probable, that traumatisms, with general concussion to the spinal cord, can exercise an exciting influence.

Symptomatology.—Depending upon the cause underlying the disease, and the extent of the pathological process in the spinal cord, i. e., of the functional injuries caused thereby, the disease picture of combined system disease may develop in various ways. The course of the disease is not rarely very acute, with rapid development of far-reaching spinal functional disturbances; in other cases, the disease shows rather a subacute course, changing in the form of its symptoms with the spread of the changes to various levels of the white matter, and in the cross-section of the white fibre system. A chronic, quite slowly progressing form of the symptom complex has also been reported. In the beginning of the disease at times there is nothing more noticeable than slight subjective disturbances of feeling, *parœsthesias:* probably, in other cases also, slight pains of the character of irritation symptoms of the roots as well, and there are cases in which more important objectively detectable spinal symptoms of functional loss are never developed. But in most instances, the system degenerations lead to clearly recognizable functional disturbances in the lateral columns, preeminently in the pyramidal tracts on the one side, and the posterior columns, probably with the inclusion of the radiation areas of the posterior roots on the other side. To these are added eventually the phenomena of functional loss, which are dependent upon a participation of the lateral cerebellar tracts and the antero-lateral columns.

Preeminently *two groups of symptoms* appear concurrently as they appear in tabes and *Friedreich's* disease for the posterior column affection, in spastic spinal paralysis for the lateral column affection. By the union of both, or

an overlapping of the groups of symptoms, there arise various types of combined system disease (*Oppenheim, E. Müller*). *Degeneration of the posterior column* alone leads to the *loss of the deep sensibility, to ataxia, to hypotonia of the limbs and to the loss of the tendon reflexes.* The degeneration of the *lateral pyramidal tracts* causes *paresis* of the *extremities*, especially in the legs with *predilection for the flexors, synergisms* (tibialis phenomenon, etc.), after longer duration a tendency to *contractures*, changes of the *cutaneous reflexes* (*Babinski, Oppenheim*), *exaggeration of the tendon reflexes*, and particularly to *hypertonia of the muscles.* If both systemic diseases occur together, the symptoms must, in part, mutually neutralize each other, according as the affection of the posterior columns or that of the lateral columns gradually and according to their special expansion preponderate. If, as is frequently the case, the functional lesion of the *pyramidal tracts* predominates in the clinical picture, the symptoms of *hypertonia*, of *spastic paraparesis*, usually, since it is a matter of disturbance in the dorsal segment, in the legs alone, with exaggeration of the tendon reflexes, synergisms and changes of the cutaneous reflexes step into the foreground, while disease of the posterior columns makes itself felt only by *slight disturbances of the deep sensibility* in the distal sections of the extremities, as well as by *ataxic* disturbances in movement, which may be increased upon participation of the cerebellar lateral tracts by the phenomena of static ataxia. Then we have the frequently observed symptom complex of *spastic ataxic paraparesis.*

If, on the other hand, the injury to the *posterior column* predominates, or gradually overshadows the lateral column lesion, the disease picture assumes a more tabetic character. The *ataxia* makes itself felt in a more considerable degree, the *sensibility disturbances* become more prominent, the *tendon reflexes disappear.* At times, even the *paresis* of the lower extremities becomes *flaccid*, the muscles become *atonic.* In most of these cases, however, some degree of *hypertonia* (while tendon reflexes are lost) persists as a sign of the simultaneously present lateral column lesion; this, beside the *change of the cutaneous reflexes* (*Babinski*, etc.), beside a slight *paresis of the predilection type* and characteristic *changes in the posture of the legs*, which finally pass over into *contractures* (dorsal flexion of the great toe, supination of the foot in the equinus position) permits us to conclude, that there is participation of the pyramidal tracts in the process of the disease.

More extensive *disturbances in sensation*, even in the realm of superficial sensation, suggest a participation of the tracts of the antero-lateral columns. It is worthy of note that the sensory disturbances in combined system diseases of the above-mentioned, more pseudosystemic form, are occasionally sharply limited upward, and so can simulate a pure cross-section lesion (*Nonne, Früind*). To the realm of the affections of the posterior columns in the wider sense belong the bladder disturbances, which are very frequently observed in cases belonging to this group: retention of the urine, frequently insurmount-

able vesical tenesmus, finally loss of the voluntary regulation of the *bladder function*. *Rectal disturbances*, retention of the feces, coprostasis occur also.

In rare cases, the degenerative process does not stop, continuing even to the higher *bulbar* divisions of the central nervous system and leads to the phenomenon of obsessional crying and laughing appertaining to the spastic symptom complex, to dysarthria, nystagmus, vertigo, and a high degree of cerebellar ataxia.

The realms of the sensory cerebral nerves do not, as a rule, participate in the process of the disease. Several times a neuritis optica has been observed to exist in the combined system disease analogously to certain forms of disseminated myelitis. The pupillary phenomena are not changed or influenced in the former.

The **diagnosis** of combined column disease is difficult and only to be made very cautiously. It can be based upon proof of the existence of the combination of posterior column symptoms with lateral column symptoms, when either the tabetic or the spastic symptom complex predominates, when signs are wholly lacking of a focal injury in the gray matter of the spinal cord (dissociated disturbances in sensation, degenerative paralyses and localized muscular atrophies) and upon the absence of symptoms of irritation pertaining to the posterior roots (relatively violent pains). The course of the disease, which usually is free from suddenly appearing exacerbations, is also, to a certain extent, a characteristic feature. It is to be remembered that in *tabes* likewise there often appears a combined system disease, a participation of the lateral columns, and in *progressive paralysis* as well; the existence of pupillary disturbances, lancinating pains, examination of the cerebro-spinal fluid will permit us to recognize under certain conditions, the tabetic nature of the changes. Furthermore, all those spinal diseases, which cause a functional disturbance of the posterior columns and the lateral columns to appear through disseminated disturbances or a slighter cross-section lesion, as cerebro-spinal syphilis, which is characterized by relatively violent pains and, occasionally, by early appearing peripheral paralyses, *compression of the spinal cord*, *spinal gliosis*, etc., are to be excluded before one is justified in thinking of a combined system disease. Differentiation from the spastic ataxic paraparesis of *multiple sclerosis* is made possible by demonstration of the purely atrophic optic affection, which is rarely absent in this disease, of the intention tremor, of the absence of the abdominal reflexes in the early stages of multiple sclerosis (*E. Müller*).

Prognosis.—The forms of system disease that have been discussed here, permit no favorable prognosis in the majority of cases. The degenerative processes, in most instances spread farther and farther and, together with the more or less quickly progressing functional limitations and the complications caused by the bladder disturbances, and eventually also by bed-sores, lead, in the course of a few years, to death. At the same time the lessening of the

general ability to resist, due to the constitutional changes which are the basis of the disease, diseases of the blood, chronic intoxications and infections have a pernicious influence. Nevertheless, the disease may possibly become stationary, and occasionally with an improvement in the general condition, a far reaching restitution, leaving only slight functional defects, may take place.

The **therapy** must, above all, take into consideration the general condition of the patient and aim at getting rid of toxic substances, at the improvement of the conditions of nutrition, and in anæmics, so far as possible, at stimulating the formation of blood. Therefore the dietetic and medicinal treatment is guided preeminently by the type of the internal disturbance. The possiblity of a syphilitic disease of the blood-vessels must not be overlooked.

In respect to the spinal disturbances, all possible care and rest is necessary for the purpose of avoiding all over-exertion of a system which is often still capable of performing its functions within moderate limits. Therefore all forced movement exercises and bath procedures are contra-indicated. On the other hand solicitous care of the skin, with careful furthering of diaphoresis is recommended; the emptying of the bladder is to be supervised and care must be taken to keep the bowels regular. After the functional disturbances have become stationary for a long time, and when there is an obvious improvement in the general strength of the patient, slowly increasing exercises may gradually be begun. Galvanization of the back along the spinal column may be recommended. In the grave progressive cases, great care, a position in bed calculated to avoid bed-sores, etc., may help in sparing the patient, who is kept in bed by ataxia and paralysis, painful suffering.

(i) Bulbar-paralytic Diseases

BY

FR. JAMIN (Erlangen)

1. Progressive Bulbar Paralysis

Paralysis Glosso-labio-pharyngea Progressiva

Progressive bulbar paralysis belongs to the systemic diseases of the motor nervous system and is closely related to the spinal diseases, which are characterized by primary destruction of the peripheral motor neuron and the clinical phenomena of paralysis and degenerative muscular wasting. Even in spinal progressive muscular atrophy frequently, and in amyotrophic lateral sclerosis almost regularly, besides the gray anterior columns of the spinal cord, the nuclei of the motor cerebral nerves which are of equal functional importance, are gradually implicated by the disease process in the form of progressive atrophy. The nuclei of the nerves of the eye muscles are an exception to this. In rare cases, for reasons hitherto unknown, the primary disturbance of the

motor system is limited from the first to these bulbar motor nuclei of the medulla oblongata and the region of the pons, and then there develops the well characterized symptom complex of a progressive paralysis and muscular atrophy in the areas of the musculature of the tongue, face, palate, pharynx and larynx, the picture of progressive bulbar paralysis.

With the exception of the rather rare atypical cases in childhood, the disease is found as a rule at a somewhat advanced age, generally near the fiftieth year, scarcely before the thirty-fifth or fortieth. Familial appearance has been very seldom reported—as a rule the cases are isolated. Yet the assumption of congenital weakness of the bulbar nuclear centers as a foundation for the disease is not unjustifiable, on the analogy of the other related systemic motor diseases and in the absence of definitely proven exogenous causes. How far one is justified in regarding colds, traumatisms, emotional disturbances as causal factors, a critical analysis of each individual case must decide. It is not improbable that the over-exertion of the injured muscular regions, especially of the muscular apparatus of the buccal cavity in certain occupations (blowing musical instruments, glass blowing, etc.), has a peculiarly injurious influence, if a diminished power of resistance is present at the same time. Possibly the diminished resistance to the demands made upon the economy by daily life, which is a sequela of syphilis and other infections or intoxications, may also be limited to these regions.

Course of the Disease and Symptomatology.—The phenomena of the disease develop very slowly and insidiously, as in progressive muscular atrophy. Step by step in the comparatively narrow field of action, one section after another is deprived of its function, in irresistable succession. Since the muscles here concerned represent, in many respects, the guardians and the assisting preparatory forces for the orifices into the gastro-intestinal canal and the respiratory passage, their functional disturbance soon causes a disturbance of the vital processes, nutrition and respiration. Therefore in every bulbar motor disturbance, the duration of the disease in comparison with that of the other systemic diseases, which affect only the locomotor capacity of the patient, is limited. Thus progressive bulbar paralysis, in spite of its usually regular, slow development, which rarely proceeds by greater strides, following one another more quickly, ends in death after a few (two to five) years by disturbances in nutrition, or by diseases of the respiratory organs.

The first signs of the disease become noticeable to the patient himself and to those surrounding him through difficulty in *speech*. The extraordinarily fine degrees of movements of the tongue, that are essential in manifold combinations for the production of sound, are already influenced at a time when the disjunction of separate contractile elements in the midst of the mighty muscular mass of the tongue, could not have altered its simple general movements and its general noticeable volume. In progressive bulbar paralysis, likewise, muscular wasting and paralysis go hand in hand;

but the functional abilities, which have been acquired with exceptional difficulty, and which are essential, especially for the complicated mechanism of speech, so influence bulbar paralyses, that the functional disturbance must be noticeable quite early, even in the slightest injury to the muscular pliability and action, and before the myoatrophic loss is visible. The composition of the sentences and the placing of words and sounds are altogether undisturbed, only pronunciation becomes difficult, confused, unclear, and thick; it is a matter of a pure *articular or dysarthric disturbance of speech.* At first, the utterance of the *lingual sounds* suffers (D, T, L, R, N, S, Sh, I) in which the participation of the *soft palate* soon makes itself felt by a nasal accessory sound and hindrance in the formation of the sounds (C, K, Ch) produced in the back part of the buccal cavity. Afterwards, the *labial sounds* are also more and more affected (B, P, F, M, W, O, U, E). The weakness of the *muscles of the larynx* which is added late, causes hoarseness, a loss of tone in the voice, monotony in a deep range, finally aphonia, so that the patient can make himself understood only with difficulty because of an inarticulate thickness of speech and stammering.

Soon after the appearance of the first speech disturbances, the first *difficulties in swallowing* become apparent. The awkward tongue is unable to direct the food which is in the mouth correctly down into the throat; the paralysis of the soft palate frequently causes a regurgitation of fluid food from the nasal cavity; solid food, because of the insufficiently functioning epiglottis and of imperfect occlusion of the glottis, reaches the larynx and the trachea; the musculature of the throat loses its ability to force the bits of food into the œsophagus.

If the patients are examined at this time of obvious functional disturbances, the bulbar *paralyses and atrophies* are usually found present.

The *facial muscles* are wasted in the lower half of the face. Especially the lips seem thin, wrinkled. They cannot be closed satisfactorily, nor can the mouth be pursed; whistling is impossible. At the same time, the muscles of the forehead and the eyelids, as well as of the eyes, as a rule remain intact and functionate. The *tongue* can be moved only with difficulty and insufficiently. It lies limp in the buccal cavity, its surface is wrinkled, crossed by deep furrows, has thin edges (Fig. 72) and displays active *fibrillary tremors*, which may also appear in the lips. The *uvula* hangs low, and is but little raised in phonation, if at all. The immobility of the *floor of the mouth* and of *the base of the tongue* is completed by the paralysis and atrophy of the muscles inserted in the hyoid bone from the innervation region of the trifacial, the facial, and the hypoglossal nerves. A laryngoscopic examination discloses the imperfect occlusion of the glottis by means of a phonation test. Active *swallowing movements* and the raising of the larynx connected with them are lost. A paralysis of the *muscles of chewing* also appears in many cases with atrophy of the temporals and the masseters

and an inability to shut the jaws properly and to make extensive lateral movements of the lower jaw.

In the affected muscles, single fibres and bundles of fibres are retained, and remain functional for some time, so that for a long time the change in electrical excitability can not be demonstrated at all, or only with difficulty. But in the atrophic tongue, in the thin pliable lips, a *partial reaction of degeneration* is frequently found, and the sluggish character of the contraction in direct galvanic stimulation is unmistakable.

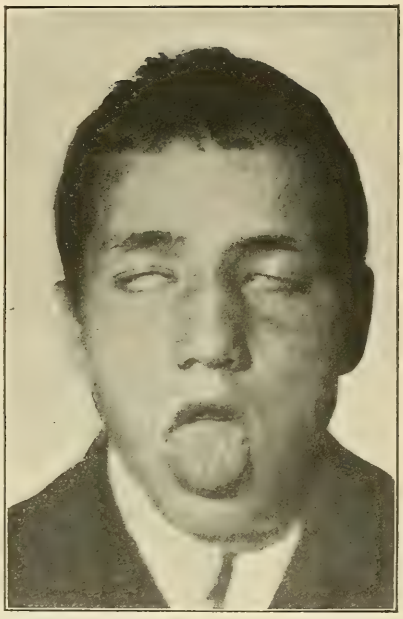


FIG. 72.—Atrophy of the tongue in infantile progressive bulbar paralysis. Flaccid features. Because of the co-affection of the upper region of the facial the lids are not perfectly closed. (*After Schoenborn and Krieger.*)

The *reflex excitability* of the atrophied muscles is, as a rule, considerably *weakened*, or even lost. In several cases, however, analogous to the phenomena of amyotrophic lateral sclerosis, obviously with simultaneous lesion of the proper central (pyramidal) tracts, an exaggeration of the reflexes in the facial region and especially of the masseter reflexes could be established. On the other hand, the palatal and throat reflexes are usually altogether lost. In "swallowing the wrong way" which is of frequent occurrence, reflexly excited coughing appears, but since an expiratory increase of pressure in the air passages is impossible because of the insufficient closing of the glottis, the hoarse coughing-up lacks effective force. Therefore foreign bodies, food and bronchial secretions can no longer be ejected, and there results the very dangerous *deglutition pneumonias*, serious bronchitis, broncho-pneumonic processes and gangrenous local lesions in the lungs.

The **sensibility** even in the area of the trigeminus is never disturbed in any respect, nor the function of the sense organs or of the sense of taste. Slight vaso-motor disturbances, rush of blood to the head with a feeling of heat, and hyperæmia occur. Observations of a considerable *increase in the pulse rate* seem to indicate that the vagus is affected.

Paralysis and muscular wasting appear first in the tongue, then in the lips and in the muscles of the lower part of the face; finally in the soft palate, the floor of the oral cavity, pharynx, larynx, and the muscles of chewing. Hence the paralysis is restricted to the hypoglossus, facialis in its lower division, motor trigeminus and vagus accessorius. Only rarely do the motor dis-

turbances spread to the muscles of the neck, the shoulder girdle and the extremities. The upper facialis area is only exceptionally affected (*Oppenheim*).

In the later stages of the disease, a profuse *flow of saliva*, which is very annoying to the patient, is nearly always observed. In part, this may be actually due to a secretory disturbance with increased production of saliva. But, on the other hand, salivation, the continuous out-flow of saliva from the corners of the half-open mouth, is certainly evoked indirectly also by the dis-

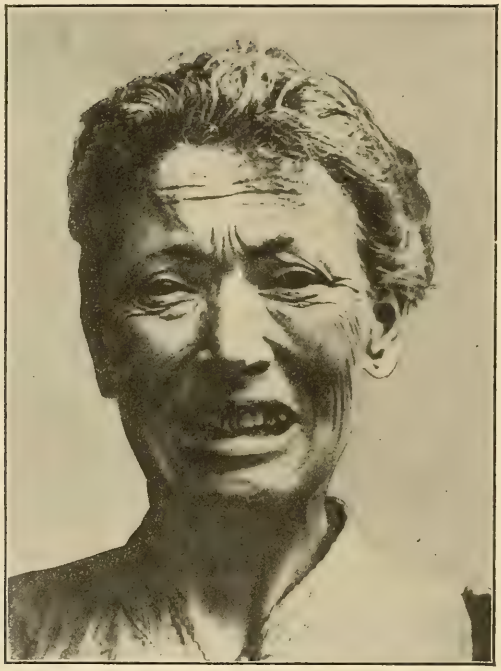


FIG. 73.—Progressive bulbar paralysis. "Transverse laughing" and impossibility of closing the lips because of atrophy of the facial muscles. The emaciated face shows folds because of the "skin having become too wide." (*After Schoenborn and Krieger.*)

turbances in movement, so that the saliva produced can no longer be swallowed.

The completely developed picture of progressive bulbar paralysis is very characteristic (Fig. 73). The face is rigid, thin, and wrinkled, only the movements of the forehead and eyes give expression to the intellectual life. The mouth is somewhat open, the lower lip hangs down and permits the saliva to escape, the corners of the mouth droop. In attempting facial expression, in crying or laughing, the mouth is drawn slightly outwards, which gives the face a distorted, suffering look. The inanition produced by the disturbance in swallowing still further increases the prominence of the morbid emaciation, since every particle of the cushion of fat is absorbed.

Pathological Anatomy.—To the selective injury of the function and nutrition of the muscles from a part of the region of the motor cerebral nerves, corresponds in the anatomic picture of the paralysis glosso-labio-pharyngea, an *atrophy* of the *nuclei of the cerebral nerves* concerned. In the nucleus of the hypoglossus, wasting and atrophy of the ganglion cells are most clearly defined, but they are found also in the nuclei of the facialis, the motor trigeminus, and the motor parts of the glosso-pharyngeus and vagus-accessorius. Instead of the wasted ganglion cells, and the network of nerve fibres, which has been for the most part destroyed in the degenerated nuclei, there appear proliferation of the glia fibres and thickening of the vessels, but without any decided inflammatory changes. The motor roots of the nuclei that have wasted, are degenerated, likewise the appertaining peripheral motor nerve fibres down into the muscles, which themselves show advanced atrophy according to the manner of the spinal muscular atrophy. In some cases, besides this, a participation of the pyramidal tracts also, just as in amyotrophic lateral sclerosis, has been found to be present. The close relations of progressive bulbar paralysis to this and to spinal muscular atrophy have been mentioned at the beginning. They are of one type, as primary selective atrophic processes of the motor nervous system, the clinical appearance of which depends only upon whether the localization of the injury is to be found in the central tracts, the bulbar or the spinal nuclei.

The **diagnosis** of progressive bulbar paralysis offers no great difficulties, if the course is considered which usually advances very gradually in typical succession from the musculature of the tongue to that of the face and pharynx, as well as the nature of the disease as a purely degenerative atrophic paralysis within the boundaries defined. Differentiation from the rest of the motor systemic diseases is founded upon the condition of the musculature of the body and the extremities, possibly upon the presence of spasms. The similarly localized paralyses in acute bulbar diseases show a different course, and scarcely ever remain so free from participation of the tracts for movement in the extremities, and for sensory conduction-tracts as they are so closely packed together in the medulla oblongata. In these, as in bulbar gliosis, multiple sclerosis, and tumors, the lesion of the cerebral nerves, especially, is more extended, not so peculiarly circumscribed, attacking also the muscles of the eye and the organs of sense. Pseudo-bulbar paralysis (cf. p. 333) is distinguished from the genuine paralysis by the swifter, frequently quite sudden or jerky progress of the disease picture, by many phenomena which suggest a diffuse participation of the brain (psychic disturbances, paresis of the extremities) and by the preservation of a far-reaching reflex ability in the paralyzed, but not limp atrophic bulbar muscles.

The **prognosis** of atrophic bulbar paralysis is absolutely unfavorable. The affection is incurable in its phenomena of nervous functional loss, and because of the disturbances in swallowing, and the insufficient protection

of the air passages, these, in turn, soon cause, as has been mentioned above, complications in the lungs and bronchial tubes; malnutrition is another important factor. Then, too, the paralysis of the larynx may lead directly to dangerous attacks of choking, and, under certain conditions, as result of a disturbance in the central regulation of the heart's activity, associated with exhaustion, sudden paralysis of the heart may appear.

Nevertheless, the pitiful condition of the patient, who with an entirely clear intellect, suffers extremely, demands great care and **treatment**. Beside the general strengthening by means of baths, nourishing foods and various restorative procedures at the beginning of the disease, an attempt should be made to strengthen the resistance of the muscles retained, in the slightly disturbed parts, by galvanization of the nuchal region, also by tonics (strychnine, nitrate of silver). As long as the cause is not entirely clear, treatment with iodide of potassium should not be omitted. At the beginning of the difficulties in swallowing, nutrition finds serious tasks. Now it is a question of reducing, so far as possible, the trouble of swallowing by a fluid diet, or one of thin porridge, with corresponding assistance; the artificial food preparations are here very useful and are still more necessary when, because of too great danger of choking, the food must be taken by means of a tube either through the nose or throat—the nasal tube being occasionally more easily inserted. The slow contractions of the atrophic pharyngeal musculature, which can be evoked by galvanization in the neck further the swallowing act, and have, at least, the good suggestive influence in that the patient can then feel the swallowing movements which he can no longer actively perform. The excessive flow of saliva can be combated by atropin. Even though the affection in itself causes no pain, still, in the last stages, the restless condition of the patient, who is tortured by difficulties in swallowing, by the violent and ineffectual cough, by difficulty in breathing, and a feeling of oppression in the heart, demands the administration of narcotics.

2. Acute (Apoplectic) Bulbar Paralysis

Phenomena of paralysis in the areas of the motor cerebral nerves after the manner of bulbar paralysis may develop in acute form also, if a sudden injury strikes the motor nuclei situated in the regions of the pons, and in the medulla, the roots or the supranuclear tracts of the cerebral nerves. These acute bulbar paralyses appear but rarely in so symmetrical an arrangement as the movement disturbances in the slowly advancing atrophic bulbar paralysis, and only in the rare cases, in which a disease corresponding to acute poliomyelitis anterior attacks the bulbar nuclear fields alone, or as bulbar infantile paralysis, together with the inflammation of the gray anterior columns of the spinal cord, do they remain wholly restricted to the functional disturbances of the motor cranial nerves. In the more frequent

lesions of the nuclei of the motor cerebral nerves due to hemorrhages, thrombotic or embolic softening, traumatic degeneration and acute inflammation in the medulla oblongata and the pons, the neighboring sensory centers and conduction tracts, the motor tracts for the extremities and the rest of the central apparatus of the medulla oblongata do not escape. According to the location and the extent of the central lesion, there are formed, in manifold combinations, complicated disease pictures, which are especially characterized by the appearance of *alternating and crossed paralyses and alternating phenomena of sensory functional loss.*

Ætiology and Pathological Anatomy.—Acute bulbar paralysis is most frequently caused by *diseases of the blood-vessels.* Arterio-sclerotic changes in the *basilar* and in the *vertebral arteries,* as well as in their end twigs, lead, through narrowing of the lumen of the arteries to insufficient blood supply of the bulbus, and sometimes even by complete thrombotic occlusion to ischæmic necrosis in the neighborhood. The branches of these arteries, especially the *arteria cerebelli inferior posterior,* which springs from the vertebralis, can to this extent be regarded as *end arteries* in that usually, when there is sudden closure and difficulty in the circulation, as a result of general atheromatosis of the cerebral arteries, sufficient collateral circulation fails to develop. The consequence of such occlusion of the blood-vessels, which in rarer cases can also be produced by an embolus from the heart, especially in the left vertebral artery, is the local death of tissue, with consequent softening and formation of cysts. Similar changes appear, also, in consequence of *syphilitic endarteritis.* In rarer cases, arteriosclerotic changes and the formation of miliary aneurisms, especially under the influence of factors that increase the pressure of the blood, cause larger focal, or small multiple hemorrhages with destruction of tissue in the substance of the pons and the medulla. Larger aneurismal dilatations of the basilar can, just as other narrowing processes do, through compression of the medulla, lead to bulbar disturbances; in their most destructive form, they are seen in fractures and dislocations of the upper two cervical vertebræ. Severe traumatisms upon the back of the head, can also give rise to hemorrhages in the region of the bulbar centers.

Besides these softenings or hemorrhagic disturbances caused by changes or injuries in the arterial system, *inflammatory encephalitic* bulbar diseases are observed, which, especially in youth, appear in connection with infectious diseases, or even in the wake of toxic processes. Perivascular infiltrations, small hemorrhages, degenerations of the nerve fibres and the ganglion cells, granular cells, proliferation of the vessels then disclose on microscopic examination a more diffuse injury to the nervous system. Lighter disturbances of this kind may be recovered from before a more deeply rooted injury of the nervous substance has occurred. Such cases exhibit but temporarily the symptomatic picture of acute bulbar paralysis; in a similar

way, but without participation of the long conducting tracts, it may be simulated also by a multiple neuritis of the bulbar nerves. We must likewise mention here, as a very rare occurrence, *abscess* of the medulla oblongata, which is usually metastatic.

Symptomatology.—The phenomena of acute bulbar paralysis appear in those cases in which they are evoked by hemorrhages or thrombotic softening, stroke-like, after the manner of a *stroke of apoplexy*. As a rule, slighter subjective complaints like headaches, especially in the back of the head, and attacks of dizziness precede for some time, or, perhaps, for a few days only, as signs of the disease of the blood-vessels, which frequently accompanies chronic nephritis, then the patient suddenly falls down with a violent attack of vertigo, associated with headache. Consciousness is often retained, but may be lost. Vomiting, tinnitus aurium, clonic twitchings, in rare cases (especially in hemorrhages of the pons), a single or even repeatedly occurring epileptoid convulsive seizures accompany the stormy outbreak of the phenomena. As a rule, the symptoms of labio-glosso-pharyngeal paralysis may then be immediately recognized. Occasionally, a few days may elapse before the picture of the disease is fully developed. Although the paralysis of the cerebral nerves and also of the extremities is at first unilateral it may, as the occlusion of the vessels progresses, seize upon the other half of the body, and an extension of the lesion upwards or downwards may be recognized by the fact, that ever more and more bulbar functions are being attacked. In cases of inflammatory bulbar affections, the development of the disease may occur also in a subacute manner, and extend over a number of days or weeks.

The articulatory *dysarthric disturbances in speech*, which appear with retained sensorium, or when consciousness returns immediately after the apoplectic attack, as a result of the *paralysis of the muscles of the tongue and larynx*, are specially characteristic. Speech becomes thick, difficult to understand, hoarse and aphonic. To this is added the early appearing *deglutition paralysis*, combined with *paralysis of the soft palate*, and the loss of the *pharyngeal and palatal reflexes*. The patients can not swallow either solid or liquid food. The latter regurgitates through the nose; frequently, also, the food goes into the windpipe, causing a troublesome cough which, because of the inefficient closure of the glottis, does not lead to sufficient expectoration. The *paralysis of the face* is still more striking. This frequently attacks all the *branches of the facialis*, causing a relaxation of the features, insufficient closure of the lids and impossibility of active or reflex movements of the facial muscles, occurring usually on one side but occasionally on both. This facial paralysis is often followed by an *abducens paralysis* on the same side, or the inability to direct vision to the side of the paralysis (cf. below). The *muscles of chewing* are not rarely attacked by the paralysis; in other cases, especially at the beginning of the disturbance, because of

central irritation due to a focus in close proximity to one of the motor nuclei of the trigeminus, trismus is observed.

According to the *location* and *extent* of the central lesion, the motor disturbance in the *cerebral nerves*, and, when the pyramidal tracts participate, in the *extremities* also, may assume various forms in various combinations (cf. Figs. 74 and 75).

If the disturbance extends cranially up to the region of the *cerebral peduncles* and the area of the *corpora quadrigemina*, the central tracts for the extremities and for the seventh, tenth and twelfth cerebral nerves, in cases of one-sided focal disease, are affected above their crossing, together with the nuclei of the oculomotor, and there appears a *hemiplegia alternans superior* (*Weber*): a paralysis of the oculomotor on the side of the lesion, and a supranuclear paralysis of the bulbar nerves and the extremities on the opposite side (Fig. 75, *II*).

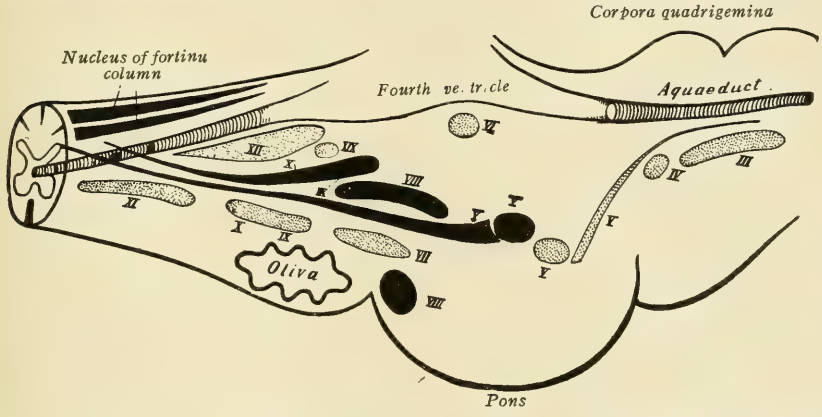


FIG. 74.—Schematic view of the location of the nuclei *III* to *XII*. Motor nuclei dotted, sensory deep black. *XIX* means *X* and *IX* (common nucleus of Nr. *IX* and *X*). (*After Villiger*).

Hemiplegia alternans inferior (*Gubler*), in which the nucleus of the facialis, or its already crossed supranuclear tract, is drawn into the focal injury, whereas the pyramidal tract is destroyed above the crossing in the pons, is more frequently found, so that a paralysis of the face, usually flaccid, is combined with a spastic paralysis in the extremities on the opposite side. At the same time there is often a paralysis of the abducens on the side of the lesion, or, if the nucleus of the abducens and its surroundings, with the connections with the nuclei of the muscles of the eye are injured, there is an associated ocular paralysis; the ability of both bulbi to turn to the side of the lesion is lost (Fig. 75, *III* and *IV*).

If the focal disease is confined to the *deeper parts of the medulla oblongata*, the unilateral lesion of the nuclei of the vagus-accessorius and of the hypoglossus or its motor roots, may strike the pyramidal tracts above the crossing,

and then there occurs a paralysis of the tongue and the muscles of swallowing, as well as of the soft palate on the side of the lesion, which, moreover, leads from the onset to severe disturbances in deglutition—and paralysis of the extremities on the opposite side (*hemiplegia alternans infima*) (Fig. 75, *V*).

Should the central injury finally strike the *pyramidal crossing* also, it may happen that the central tract for the upper extremity is interrupted above the crossing, while that for the lower extremity may be affected below the crossing; then the arm on the side of the lesion and the leg on the opposite side, will be paralyzed, i. e., exhibit a spastic paresis (*hemiplegia cruciata*) or paralysis of both legs may appear or, more rarely, of both arms.

Since, owing to the small size of the pons and the medulla, the softening and inflammatory foci frequently pass beyond the middle line, and as the boundaries of the foci are, as a rule, irregular, there occur also bilateral disturbances, paralysis of all the extremities, combined with unilateral bulbar nuclear paralysis in the facialis or of the oral or pharyngeal cavities, or even bilateral glosso-labio-pharyngeal paralysis, connected with unilateral paresis of the extremities.

The motor tracts for the *extremities* are, in bulbar focal lesions, always affected above the spinal motor nuclei; the paralysis of the extremities, therefore, corresponds in its details to cerebral hemiplegia; if the disease continues long enough, there develops a *spastic paresis of the extremities*. Not infrequently, however, the tendon reflexes are lost, especially in the upper extremities, probably as a result of synchronous lesion of the spino-cerebellar tracts; the Babinski reflex also does not appear regularly in bulbar paralysis.

The *paralysis in the areas of the cranial nerves* is in most cases of acute bulbar paralysis, throughout, or in the greater part of the paralyzed region, *degenerative, flaccid*, corresponding to the lesion of the peripheral motor neuron in the nuclear area, or in the motor roots. This may be demonstrated in the individual case by the *absence of reflex excitability in the musculature of the face, tongue, and throat.* If the paralysis persists for several days, the presence of a quickly *progressing atrophy* in the paralyzed facial muscles and in the tongue, as well as *fibrillary twitchings* and *electric reactions of degeneration* may be seen to develop in many cases. The latter may occasionally be evoked in the musculature of the pharynx also.

Next to the motor disturbances which are mostly of an alternating character, the condition of *sensation* in acute bulbar paralysis deserves attention. Here, also it is to be remembered that the conduction of the sensibility of the trunk and the extremities, undergoes, for the most part, a *decussation* in the higher parts of the medulla oblongata, so that with a focus that appears on one side in the pons, the sensory tracts of the trunk and the extremities may be affected on the opposite side, in addition to the sensory roots and centers of the trigeminus on the same side of the lesion; this would result in a *hemianæsthesia cruciata*, with paralysis of sensation in the trunk

and the extremities on the opposite side and loss of sensation in the face (especially in the second and third branch of the trigeminus) and in the oral and pharyngeal cavities on the side of the focal injury. In the deeper divisions of the medulla oblongata (olivary region) relations are complicated by

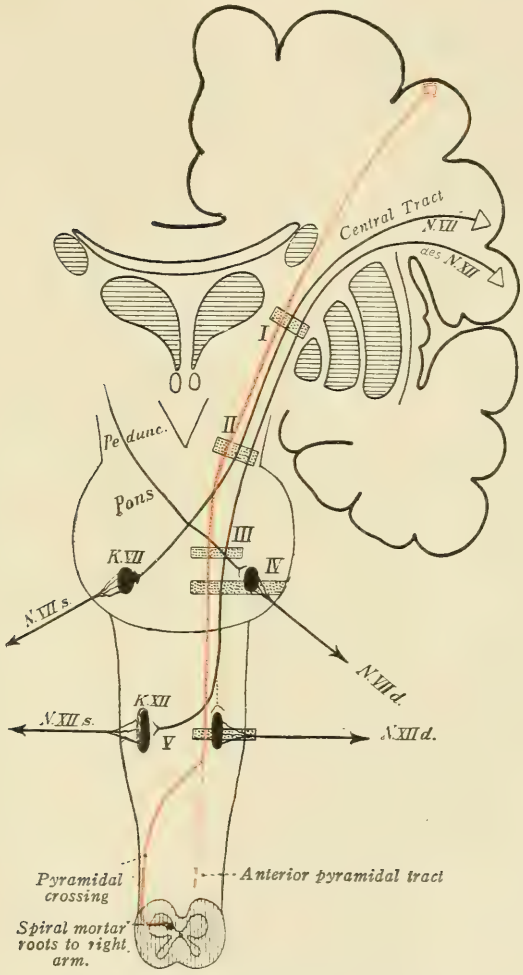


FIG. 75.—Scheme of explanation of hemiplegia alternans (for the seventh and twelfth nerves). Red: pyramidal tract; *N.VII s*, left facial nerve; *N.XII d*, right hypoglossal nerve. *I*, focus in the right inner capsule left half of the body, the left seventh and twelfth nerves affected; *II*, focus in the right peduncle, the same result; *III*, focus in the right half of the pons, left part of the body and the left twelfth nerve, the right seventh nerve (supranuclearly) affected; *IV*, the same caudad, left part of the body, left half of the tongue and the right seventh nerve affected (nuclearly resp. radicularly); *V*, a focus in the medulla paralyzes the left half of the body and the right half of the tongue (nuclearly or radicularly). (*After Liepman.*)

the fact that in near vicinity of each other, three sensory systems of various importance are situated: 1. In the medial division, near the raphe (interolivary layer) the *kinæsthetic tracts* (sensation of position, muscle sense, pressure sense—deep sensibility), which are crossed here, by the aid of the

fibræ arcutæ internæ. A focus situated higher up will therefore cause a disturbance of the deep sensibility on the opposite side, a lower focus may be followed by a destruction of the deep sensibility on the side of the lesion. 2. A lesion in the lateral division (tractus spinotectales and spinothalamicæ), the *tracts for the sensations of pain, of cold, of heat,* which have crossed already in the spinal cord, brings with it a dissociation disturbance of the sensations of pain and temperature on the side of the body opposite to the focal injury (trunk and extremities). 3. The spinal root of the trigeminus with the substantia gelatinosa, which is the medium for *conduction of sensibility for the face,* especially in the region of the first branch of the trigeminus, in all its qualities. Since the fibres of the trigeminus, which serve for pain and temperature sensation, are crossed already in the lower sections of the medulla oblongata, a focal injury here may, according to its extent, cause a disturbance in sensibility of the trigeminus in all its qualities on the side of the lesion, or, it may produce an analgesia and thermanæsthesia on the opposite side of the face (especially in the region of the first branch of the tri-facial) (*Wallenberg, Oppenheim, Breuer-Marburg, L. R. Müller, E. Müller*).

These topographical relations explain the fact, that in acute bulbar paralysis, as a result of focal disease in the medulla oblongata, the hemianæsthesia alternans or cruciata frequently appears in the form of a *dissociated paralysis of sensation.* The sensation of touch alone, which can probably be conducted by various tracts, is rarely affected. Frequently there is a loss of the sensations of pain and temperature in the trunk and the extremities on *the side opposite the lesion,* which may, however, as we said above, extend also to the upper half of the face thus forming a *total hemianalgesia,* and which, as a result of the lesion of the long trigeminus root, is connected with a disturbance in sensation of the face on the side of the lesion. Since a hypæsthesia of the sensation of touch, but more frequently, a loss of deep sensibility on the side of the lesion, may be connected with it, there result disease pictures, which have a certain similarity to the *Brown-Séquard* type of unilateral spinal lesion, so that, in connection with the motor disturbance of the cerebral nerves, which appears on the side of the lesion, especially in the facialis, one may speak in a similar sense, of a *unilateral bulbar lesion.*

A peculiar type in this respect may be recognized in acute bulbar paralysis, which has its origin in thrombotic occlusion of the *arteria cerebelli inferior,* a branch of the vertebral. Here there is a circumscribed sharply defined softening in the lateral section of the medulla oblongata between the olive, the fibræ arcuatæ internæ and the restiform bodies, while the pyramids, the lemniscus and the floor of the fourth ventricle are often spared; the tracts already crossed here, for the conduction of the pain and temperature sensations, the (motor) nucleus ambiguus vagi, the root of the hypoglossus, the spinal trigeminus root and the restiform bodies are affected. The symptoms

noted correspond to such a lesion: contralateral hemianalgesia, unilateral paralysis of the musculature concerned in swallowing and of that of the palate, larynx and tongue, trigeminus anæsthesia on the side of the lesion and ataxia of the extremities on the same side.

Sometimes, subjective sensory disturbances, such as paræsthesias and perverse temperature sensations (cold is felt as heat, heat as cold) make themselves felt in the anæsthetic or hypalgetic extremities in acute bulbar paralysis. Homolateral hyperæsthesia has been also observed with the contralateral analgesia, just as in the unilateral spinal lesion.

Ataxic disturbances in the extremities may be founded upon a lesion of the kinæsthetic tracts, with corresponding loss of deep sensibility. They appear more frequently in the form of cerebellar ataxia, associated with phenomena of vertigo and falling to one side, in *lesions of the restiform bodies*, or the tractus spino-cerebellaris lateralis on the side of the focal injury, and then they usually remain confined to a disturbance in co-ordination in the large joints, or, say, the musculature of the trunk. But the ataxia, combined with hypotonia, may be so severe as to simulate, at first, a hemiplegic paralysis. *Nystagmus* (rotatorius), too, is frequently observed as sign of a disturbance in the co-ordination of the muscles of the eye. Disturbances in the special sensory organs rarely occur. In focal injuries that are situated very high, there sometimes appear also disturbances in sight and pupillary phenomena, occasionally, likewise, *disturbances in hearing*, when the nuclei of the auditory nerve are affected. Participation of the vestibular nuclei, or of the appertaining root fibres, then also of the cerebellum and its connection with the pons, bring about further disturbances in the harmonic co-operation of the muscles, conditions of vertigo, disturbances in equilibrium. The disturbance is always more marked on the side of the lesion.

Important are the changes which point to a participation of the nervous centers, which, located in the medulla oblongata, control the *vegetative* functions, and which often quickly cause an unfavorable turn in the course of the disease. As such, we frequently find *disturbances in respiration*, dyspnœa and especially periodic breathing, which frequently continues for a long time (Cheyne-Stokes) with occasional alarming pauses in respiration. Furthermore, *acceleration* or *irregularity* of the *pulse* and an increase in the temperature of the body, which, especially when the outcome is fatal, rises considerably (even as high as 106° F.). *Albuminuria and glycosuria* occasionally accompany the disease picture. More frequently one observes a *flow of saliva*. Vaso-motor disturbances and disturbances in the *secretion of perspiration* occur also.

That acute bulbar disease is capable of destroying also the central connections with the *sympathetic*, is shown by observation of the occurrence of Horner's oculo-pupillary symptom complex (ptosis, enophthalmos and miosis, as a consequence of paresis of the sympathetic) on the side of the

central focus (*E. Müller*). *Bladder and rectal disturbances* (retention of urine and feces) are, as a rule, present at the beginning of the disease. Priapism, likewise, has been observed in connection with the first bulbar attack.

Course of the Disease and Prognosis.—In many cases the patients die soon after the first attack; large hemorrhages, many small multiple hemorrhages and occlusion of the basilar artery lead, by injury of the important vital bulbar centers, inevitably to death. If the immediate consequences of bulbar apoplexy are overcome, life is still threatened by the results of the paralysis of deglutition, which appear as aspiration pneumonia and bronchitis and cannot be overcome, because of the insufficient nutrition and the lowered respiratory function. But in some cases, after some time, through absorption of the hemorrhages and restitution of the circulation in the injured parts, the phenomena disappear to a greater or less extent, and in time there may be sufficient restriction of the anatomic and functional defect, so that only slight motor disturbance remains. In slight inflammatory affections of the bulb, even complete recovery may occur in young patients, in spite of the phenomena of extensive functional loss in the cerebral nerves and the extremities at the beginning. If the paralyses persist, the phenomena of degenerative atrophy develop more and more distinctly in the regions of the nuclei that have been attacked (e. g., unilateral wasting of the tongue) and also the spastic phenomena in the hemiparetic extremities. Nevertheless, at the beginning of the disease the prognosis is dependent on the extent of the focal phenomena and on the severity of the general symptoms, especially the impairment of the vegetative functions.

The *diagnosis* is based on the *sudden* beginning, or at least on the unexpected onset associated with a rapid increase of the symptoms and on the arrangement of these symptoms, especially the combination of *glosso-labio-pharyngeal paralysis* with *alternating hemiplegia and alternating hemianalgesia*. From *pseudo-bulbar paralysis*, acute bulbar paralysis is distinguished by the course and the behavior of the reflex excitability in the cerebral nerves. Transitional forms, however, occur.

New growths in the medulla oblongata or in its vicinity often induce, long before the focal symptoms can be recognized, symptoms of general brain pressure; the same is true of cysticerci and chronic meningitic processes; thereby they show a quite different course, than acute bulbar paralysis, in which the distal effects are far less apparent than the focal phenomena, which can be determined with great exactness. *Bulbar gliosis* also can usually be excluded without any difficulty, by considering the development of the symptoms; it must be remembered, however, that occasionally through intercurrent hemorrhages in the medulla oblongata, an acute bulbar paralysis may be directly induced in the course of it. In *multiple sclerosis*, the pons and medulla oblongata seldom remain intact. In this

disease, the total disjunction of the bulbar centers with the corresponding striking and well-defined paralyses and disturbances in sensation, etc., never develops as quickly as in bulbar paralysis; nay more, for a long time, only the lighter bulbar functional disturbances appear, together with a number of characteristic spinal and cerebral disturbances, so that practically there can scarcely ever be a question of confusion of the two, if a thorough examination is made.

Therapy.—Treatment of a bulbar apoplectic attack is to be directed along similar fundamental principles, as that of a cerebral apoplexy. Absolute physical and psychical rest is, of course, essential; in convulsive attacks, it may be attained by narcotics. Local or general blood-letting is to be considered only when hemorrhages or local inflammation are suspected. The possibility of a syphilitic ætiology must be taken into consideration early and treatment by iodide of potassium instituted; in case of disturbances in swallowing, injections of iodipin are always advisable. Nutrition must be carefully watched over and when the paralysis of swallowing is severe, is most satisfactorily attained from the beginning by means of a stomach tube, since in this way, too, the frequent faulty swallowing and therewith the fear of aspiration pneumonia may be avoided. In unilateral vagus paralysis the patients are able, after some time, to swallow more easily again, if they incline their head towards the unparalyzed side. Cold food is more easily swallowed than warm (crossed hemithermanæsthesia). The analgesic disturbances demand at an early date careful protection of the patient in bed (water cushion) and marked care of the skin, in order to avoid bed-sores. Protection of the conjunctival scleræ and the cornea in total facial paralysis and trifacial anæsthesia must be especially considered. The use of the constant current may assist in reviving the weakened area of the motor cerebral nerves; galvanization at the nape of the neck is especially to be recommended with the anode and excitation of the movements of deglutition. If the paralyses prove after longer observation to be constant and irreparable, with unclouded consciousness, there is no sense in withholding from the severely suffering patient, the beneficent action of effective narcotics.

3. Pseudo-bulbar Paralysis

The bulbar-peripheral centers of the motor cerebral nerves subserve as do the spinal motor centers of the gray anterior columns, the conduction of cortico-fugal motor fibre systems coming from the corresponding centers of the psychomotor region of the central convolutions and in the greater part conducted through the pyramidal tracts. They transmit the *motor impulses* from the *cerebral cortex* to the bulbar nuclei and thereby, for the most part, bring about the production of voluntary movements in the appertaining

muscular regions and especially in the extraordinarily fine shadings in the movements of the lips, pharynx and larynx that are needed, for instance, in speech. *Purely reflex movements*, partly in rather complicated movement complexes (sucking reflexes), can also be produced in the bulbar centers alone, as the experience in the malformations caused by arrested development in the cerebrum teaches us. Then, too, the *involuntary movements of expression and emotion*, as in laughing and crying can be evoked, independent of the influence of the cerebral cortex, by the aid of the subcortical centers in the lenticular nucleus and the optic thalamus. All these reflex movements, which in early childhood are, in part at least, of great physiological importance, are in the older child and in the adult subordinated to the restraining, or stimulating control and the inhibitory influence of the cortex of the brain; in part, they are completely crowded out by the preponderance of cortical influence under normal conditions. A destruction or far-reaching restriction of this cortico-bulbar influence by interruption of the supranuclear, cortico-bulbar tracts, or injury of the corresponding centers of the cortex, will therefore be followed in the region of the motor cerebral nerves, especially in the musculature of swallowing, of the lips, tongue and larynx, by a *paralysis* or *paresis* in the sense of the loss of the *ability* for *voluntary movement* without marked atrophy, while reflex excitability is retained and under certain conditions also involuntary emotional and expressional movements. This is the type of pseudo-bulbar paralysis in contra-distinction to acute bulbar paralysis and to progressive bulbar paralysis, both of which, though differing in course, location and extent of lesion, are characterized by the fact, that the motor nuclei in the bulb are themselves injured and at the same time all the motor bulbar functions, even the reflex and automatic ones, corresponding to the localization of the disease, are destroyed, with the natural consequence of a degenerative atrophy, if the duration of the disease is sufficiently long.

It is significant, for the pathological physiology of pseudo-bulbar paralysis, that in the most common form of supranuclear motor disturbance, cerebral hemiplegia, there are, as a rule, no symptoms of bulbar paralysis; it is true there frequently appears a contra-lateral one-sided paresis of the lower (oral) part of the facial nerve and of the tongue, associated with the paralysis of the extremities; but the musculature of the pharynx and larynx is spared. The reason for this is that these muscles receive *bilateral* cortical innervation, and that the voluntary movements of these regions on *both* sides can be sufficiently supplied from *one* hemisphere alone. The cortico-bulbar tracts must therefore be injured or interrupted in *both* hemispheres, if there is to be a clinically recognizable supranuclear paralysis of the pharyngeal and laryngeal muscles. The conditions are similar as regards the masticatory muscles, those of the forehead, and those that close the eyelids.

Ætiology and Pathological Anatomy.—Pseudo-bulbar paralysis occurs

with relative frequency in childhood, associated with more or less prominent spastic-diplegic phenomena, in those cases in which an intrauterine or a later acquired inflammatory or traumatic injury caused a bilateral extensive lesion of the cortex of the brain, and thereby led to a functional disturbance of the psycho-motor cortical fields of the bulbar nerves as they are situated within a narrow radius.

In adult life, it is principally *arterio-sclerotic* changes of the blood-vessels of the brain, that lead to multiple hemorrhages from miliary aneurisms, to numerous thrombotic softenings, and to extensive degenerative processes in the hemispheres, and thereby to bilateral lesions of the cortico-bulbar motor systems as well as to the other disturbances. More extensive apoplexies appearing consecutively, in both halves of the brain, may have the same effect. Multiple inflammatory foci, scleroses and embolic softenings, occur less frequently in connection with this condition. *Syphilis* with its changes in the blood-vessels and gummatous focal disturbances is quite frequently a cause of extensive cerebral disease.

The location of the injury may vary. The focal disease is rarely restricted to those cortical fields, which on both sides direct the psycho-motor stimulation of the bulbar nerves in the *operculum*. More frequently, softening foci, cysts, etc., may be demonstrated in the *medullary layer of the hemispheres*, especially in the posterior parts of the frontal brain or even lower down in the radiation of medullary fibres, the inner capsule—in the region of the great basal ganglia. The cortical lesion of *one* side may combine with a focus in the subcortical medullary layer of the other side and it is characteristic of the disease that is based mainly upon disturbances in circulation, that the arrangement of the changes in the brain may vary greatly.

In some cases, symptoms of pseudo-bulbar paralysis have been observed also in merely one-sided disease of the brain. It is a matter, then, usually of very severe lesions, extensive hemorrhages, etc., in which a functional injury of the other hemisphere also seems understandable, considering the disturbances in pressure and circulation.

If the atheromatosis of the cerebral arteries has progressed so far that it leads to multiple or extensive focal changes in both hemispheres of the cerebrum, it does not seem strange that the more delicate branches of the vessels in the brain stem and in the medulla oblongata also fail to remain intact for any length of time and themselves cause changes and functional disturbances in the nuclei of the cerebral nerves, beside the cerebral lesion of the supranuclear system. We have, then, a combination of pseudo-bulbar paralysis with phenomena of acute genuine bulbar paralysis, a disease picture, that may have some traits of both forms, *cerebro-bulbar glosso-pharyngo-labial paralysis* (*Oppenheim*).

Symptomatology.—The cerebral glosso-labio-pharyngeal paralysis is

but rarely ushered in by a severe stroke. More frequently there is observed a subacute progressive development of the disease in starts or jerks, or the symptom complex is completely developed only after *several successive apoplectic attacks*. Thus it may happen that after a first attack, a hemiplegia appears with slight disturbances in speech and swallowing, the latter two disappearing, while the hemiplegia persists and that only after a second stroke which now paralyzes the other half of the body which thus far had remained intact, associated with the now bilateral injury to the brain, cerebral-bulbar paralysis becomes distinct. In other cases again, the disease may develop gradually through the slow accumulation of small, multiple foci of cerebral softening, possibly hastened occasionally by slight apoplectiform attacks, or even without noticeable phenomena.

It is characteristic of the anatomic processes, which, in connection with diseases of the blood-vessels, underlie the functional disturbances, that the phenomena of functional loss in pseudo-bulbar paralysis are not strictly confined to a definite topographical arrangement; therefore, they do not usually appear in strict symmetry and may be complicated by manifold focal or diffuse cerebral disturbances.

Especially characteristic are the disturbances in the *facial muscles* and in those concerned with *chewing* and *swallowing*, likewise in the musculature of the *tongue*, *palate*, and *larynx* with the prominent phenomena of *dysarthria and dysphagia*, which, as a rule, are combined with *general motor disturbances*.

The *exterior appearance* of the patient is striking: the face is rigid, immobile, the mouth half open, the expression lifeless and stupid. The continuous *flow of saliva* has a still more disfiguring effect. The posture of the body is stiff and awkward, the head bowed forward, the movements slow, clumsy and spastic, the gait a laborious drag with small tripping steps.

The voluntary movements of the facial muscles, of the muscles of the eye and tongue and of the chewing muscles, are very markedly limited, or entirely lost. *Speech* becomes difficult to understand, nasal, or thick, due to the disturbance of articulation and of the formation of labial sounds and to the paralysis of the soft palate. It is affected likewise in intonation, by paresis of the laryngeal muscles and disturbances in the mechanism of breathing. The difficulties in deglutition but seldom attain threatening proportions, but there is frequently a regurgitation of liquid food through the nose and solid food frequently gets into the windpipe.

The muscles, that have thus been withdrawn from the dominion of the will do not suffer serious atrophy, as a rule, even after prolonged continuation of the disease, the lips and tongue do not grow thin, show *no fibrillary twitchings* and neither upon indirect or direct stimulation do they give *electric reactions of degeneration*.

The muscular areas that voluntarily can not be set in motion at all, or, only to a very slight degree, are set into very lively, morbidly *increased* activity under the influence of emotional disturbances, or *reflexly* by cutaneous and sensory stimulations.

Thus the eyeballs, remaining ordinarily in rigid immobility, turn involuntarily in the direction of an acustic or strong optic stimulation, and the eyeballs, which can scarcely be moved at all by the patient, are able to follow a moving object by reflex adjustment (*Wernicke, Oppenheim, Strümpell*). Very slight causes evoke *spasms of crying and laughing*, in swift, often alternating succession (*Oppenheim and Siemerling*). At the same time the muscles, which are almost entirely paralyzed for voluntary movements, contract strongly; the face, formerly immobile, is now spasmodically distorted; spasmodic movements of expiration and coughing, occasionally a temporary asphyctic stoppage of breath with pronounced cyanosis of the face, accompany these peculiar emotional explosions, in which the more intelligent patients are quite conscious of the insufficient psychological motivation of these more reflex emotional and expressional disturbances, which really are a result of insufficient cerebral inhibition.

A permanent increase of tension in the paretic muscles of the face, the tongue, and the jaws (trismus), is occasionally present, and likewise irritation phenomena, like spasms in the muscles of the jaws, gnashing of the teeth, a spasmodic clearing of the throat and choking may be observed. The insufficient cerebral restraint and inhibition is further made manifest in an *increase in the reflex excitability* of the bulbar nerves and in the appearance of combined reflex movements, which under normal conditions, early in extrauterine life give way to the voluntary use of the labio-glosso-pharyngeal musculature. *The masseter reflexes* are frequently exaggerated, and upon tapping the lower half of the face, reflex contractions appear in the orbicularis oris, recalling sucking movements (*Toulouse and Vurpas*). The palatal and pharyngeal reflexes are retained. Irritation of the hard palate causes muscular contractions of the face in the cheeks or upper lip (*Henneberg's* hard palate reflex). Irritation of the lips or of the oral cavity evokes rhythmic movements of the lips, tongue, lower jaw, and in the throat (*eating reflex of Oppenhein*) in peculiar, seemingly almost co-ordinated purposeful combination. Movements of swallowing, even when there is disturbance in consciousness, may, as a rule, be evoked by the introduction of liquid food. These reflexes appear most actively in infantile pseudo-bulbar paralysis, but they are frequently found in adults also. One must notice, here, that just as in cerebral hemiplegia, the reflex increase develops to its greatest height only when the lesion has persisted for some time, and the subcortical or bulbar centers, after the interruption of the cerebral influence have, to some extent, recovered, or again become accustomed to independent activity.

That the control of *respiratory innervation* may suffer also is proven, not only by the cases of the disturbances in breathing in obsessional laughing and crying, but by the frequently occurring conditions of *dyspnœa* and *periodic breathing* (Cheyne-Stokes) and the difficulty of regulating breathing during speaking and swallowing (*Hartmann*). *Acceleration and irregularities of the heart's action* also occur. In greater impairment of the automatic functions of the bulbar centers (increase in temperature, glycosuria, etc.), however, one must consider that the arterio-sclerotic disease may likewise occasion foci in the pons and the medulla oblongata, in addition to the cerebral injuries, whereby a disease picture develops, which no longer exactly fits the frame of supranuclear bulbar paresis.

As a rule, in pseudo-bulbar paralysis, there are also *spastic motor disturbances* in both arms and legs, corresponding to the bilateral cerebral injury, or, according to the localization of the cerebral foci, only in the form of a cerebral hemiplegia. In extensive arterio-sclerosis of the nervous system, the spastic character of the paresis of the extremities may be more or less obliterated or concealed by injury of the compensation tracts and of the long and short reflex arcs. Occasionally, too, in a restriction of the central disease to the centers and supranuclear tracts of the motor cerebral nerves, there appears a pseudo-bulbar paralysis with but very slight movement disturbances in the limbs (senile abasia) (*Naunyn*) or without any at all.

Of other complications of cerebral labio-glosso-pharyngeal paralysis, we must lay stress upon the very frequent *psychic disturbances*, which make themselves manifest especially as intellectual weakness, deterioration in the ability to receive and retain new impressions, and in memory, and may be increased to disturbances in orientation, clouded intellect, conditions of delirious excitement. Of course one must beware of deciding on the presence of a psychic defect, from the external aspect alone—from the combination of a staring expression in the face with an open mouth, dysarthric speech disturbances and general difficulty in movement. Furthermore, the disease picture may be variously enlarged by a series of cerebral disturbances corresponding to the extension of the injury in the brain: motor and sensory aphasia, hemianopsia and other central visual and auditory disturbances, disturbances in equilibrium, as well as more or less extensive disturbances in sensation, especially of the sense of touch and localization, but also hemianæsthesia and disturbances in co-ordination. Among motor symptoms, athetosis and chorea from central irritation either on one or both sides are sometimes seen. Pupillary disturbances and optic atrophy have been occasionally observed along with the bulbar symptoms of functional loss.

More frequently and especially in the beginning of the disease, but also as a permanent concomitant phenomenon, pseudo-bulbar paralysis

causes a loss of the voluntary control over the *functions of the bladder and rectum*, which is present chiefly in the form of retention of feces and urine, but may lead also to enuresis and coprostasis.

The **diagnosis** of pseudo-bulbar paralysis will offer no great difficulties if there is complete development of the supranuclear bulbar paralysis with retained muscular nutrition and electrical excitability, with normal, even increased reflex and emotional motor excitability and with the frequent complications of psychic disturbances and cerebral paralysis of the extremities. A confusion with *progressive bulbar paralysis* is excluded by the fact that this has a far more insidious course, and is distinguished by degenerative atrophy, fibrillary tremors and loss of electrical excitability, as well as of the reflexes of the motor cerebral nerves and by the strict confinement to these areas with the exception of the nerves of the eye muscles. *Acute apoplectic bulbar paralysis* may occasionally be mistaken for the pseudo-bulbar paralysis, but can usually be distinguished from it by the presence of genuine bulbar focal symptoms, especially if alternating hemiplegia and alternating hemianalgesia exist. *Amyotrophic lateral sclerosis* with bulbar localization can give rise to disturbances in movement, which resemble closely those of pseudo-bulbar paralysis, but it will make itself known by the steadily progressing course without apoplectic advances by fits and starts, by the occurrence of degenerative, localized muscular atrophies and by sparing of the psyche and all functions not purely motor. In *myasthenic bulbar paralysis*, the phenomena of fatigue, weakness and exhaustion, beside the electro-diagnostic signs, predominate in the disease picture, and it wholly lacks the phenomena, so characteristic of pseudo-bulbar paralysis, of involuntary motor processes excitable by reflexes, frequently increased to veritable paroxysms in the paretic bulbar muscles.

The **prognosis** of pseudo-bulbar paralysis is unfavorable, even when one considers only the excitant factor, the atheromatosis of the cerebral arteries. The recurrence of apoplectic seizures and therewith, a dangerous turn in the disease, is always to be feared, while thanks to the reflex activity of the bulbar centers, there is usually no danger to life directly from disturbance in deglutition and aspiration of foreign bodies. With careful treatment, avoidance of all influences, that might suddenly increase the pressure of the blood (excitement, alcohol, caffein) the patients may, under certain conditions, be kept alive for a comparatively long time; with careful attention even an improvement in the disturbances is possible. *Treatment by iodide of potassium* is always to be recommended; if a syphilitic ætiology is suspected, or proved by anamnestic or objective signs (scars, *Wasserman's* serum reaction, etc.), a well-directed combined treatment of iodide of potassium and mercury may cause a considerable improvement in the disturbance and in any case effectively combat the further spread of the disease in the cerebrum and possibly in the bulb as well.

3. MULTIPLE INSULAR SCLEROSIS. (Disseminated Sclerosis)

BY

HERMANN SCHLESINGER (Vienna)

Ætiological Factors

The disease is traced back by a number of authors to early (embryological) disturbances and a disposition upon the part of the neurogliar tissue to proliferate (*Strümpell*, *Mueller*). I am inclined to agree with *Oppenheim* that congenital anomalies or such developmental disturbances as begin very early in life represent only a factor favoring the special disposition for the disease.

Marie is of the opinion that acute infectious diseases represent probably the most important ætiological factor. The same view is held by many of the later writers, as *Kahler* and *Pick* (even before *Marie*), *Oppenheim*, *Maixner*, *Redlich*, *Spiller* and others. From my own experience I should regard this as the cardinal, the causal factor.

The diseases concerned chiefly are typhoid, pneumonia, diphtheria, influenza, scarlatina, variola. Malaria also has been observed rather frequently as an ætiological factor. Intoxications (mercury, carbonic oxide, tin, possibly manganese also), may call forth the disease.

The onset of the disease has frequently been observed after severe psychic disturbances and especially after colds; furthermore also after severe traumatisms (*Schlagenhaufer*, *Mendel*, *Redlich*). In a great many cases no ætiological factors can be discovered. The period of adolescence is particularly predisposed to the disease.

Anatomic Changes

Disseminated sclerosis is characterized by the appearance of multiple lesions in the brain and spinal cord. The number of foci may range from a few up to several hundred. Their distribution is not regular; sometimes with a relatively large number of foci in the spinal cord, there are only a few in the brain. They may affect every section of the central nervous system, are occasionally arranged with striking symmetry and show quite considerable variations in size. The largest foci are found in the pons and in the brain. The optic nerve and the chiasma, also, often have one or more foci.

The foci themselves are, as a rule, very strictly circumscribed and even macroscopically are clearly distinguished from their surroundings by their gray color. They are recognizable, if situated near the surface, even before cutting into the spinal cord, and may be clearly felt by the palpating finger as harder spots. They may lie partly in the white and partly in the gray matter.

In the foci, one finds upon microscopic examination no, or only a few nerve fibres retaining their medullary sheath. But many well-preserved axis cylinders pass through the foci. The neurogliar tissue is extraordinarily increased, and forms a thick felt-like layer, which does not show any tendency to softening anywhere. The ganglion cells, even in the center of the foci, are not injured, but the vessels often show grave changes; their walls are thickened and the lumen narrowed, or even entirely obliterated. Secondary degenerations are absent, even after long continuation of the disease, and with numerous foci. Upon the advent of diffuse sclerosis, changes in the form of the sections attacked, especially in the pons, occasionally occur.

Besides the old foci, one frequently sees diseased spots in which the process is of more recent date; small-celled infiltration of the vessels (filling of the adventitial lymphatic spaces with fatty granular cells) is exhibited; processes of breaking down in the focal regions are still going on. Foci of more recent date are found especially in cases with an acute and subacute course.

The *pathogenesis* is by no means clear. The various views are, in part, in direct contrast with one another. Some authors, like *Schmaus* and *E. Müller* speak of congenital conditions (hypoplastic conditions of the nervous system). Other investigators consider proliferation of the neuroglia the primary change (*Rossolimo, Strümpell, Probst*). Still others believe in a primary lesion of the parenchymatous parts of the nervous system (*Redlich, Köppen*). The disturbance of the vascular apparatus is considered as a causal factor especially by *Taylor, Rindfleisch, Ribbert.* The theory of an inflammatory origin finds many supporters (*Charcot, Erb, Leyden, Oppenheim*). It is very probable that an acute disseminated encephalomyelitis, and in particular that form accompanied by cellular infiltration (*Oppenheim*), leaves in its wake, as a resulting condition, a multiple sclerosis.

Symptoms and Course

We are indebted to an excellent study by *Charcot* for our knowledge of several cardinal symptoms of multiple sclerosis, without which for a long time one could scarcely venture to make a strict diagnosis of this affection. Not for some time afterwards were quite a number of other symptoms discovered. In the early stages of the disease cerebral or spinal symptoms may occupy the foreground. Very frequently, the disturbances begin with *visual disturbances* as has been pointed out particularly by *Oppenheim, Bruns, Stoelting and Uhthoff.* Ophthalmoscopic examination then discloses a neuritis, or an optic atrophy.

These symptoms may be the first ("like vanguards") (*Oppenheim*), and are therefore very often misinterpreted. Since our knowledge of multiple sclerosis has been steadily increasing, it happens more and more frequently, that a "retro-bulbar neuritis" afterwards turns out to have been a precursor

of multiple sclerosis. I have seen quite a few patients, in which this was the first symptom of the disease; occasionally it precedes the other phenomena by years. In other cases, a uni- or bilateral optic atrophy develops very slowly or very quickly, with a peculiar gray discoloration of the disc: this optic atrophy as a rule is partial (*Uhthoff*), more frequently temporal than nasal, rarely ever total, and was found in almost half the cases. Since the visual disturbances in the great majority of cases are transient and are not associated with pupillary disturbances, they are frequently regarded as hysterical precisely because of their instability, when there has been no examination of the fundus. *Bruns-Stoelting* places the frequency of cases beginning with optic atrophy at 30%. Even very marked visual disturbances may disappear. One of my patients, who, five years ago, was unable to recognize fingers, now reads the finest print without difficulty.

Paralyses of the muscles of the eye, especially of those supplied by the oculomotor and the abducens, are frequent in the early stage. Ophthalmoplegia is very rare—as a rule, only a few of the muscles are disturbed and the internal muscles of the eye remain normal. Often the paralysis affects the associated movements. Permanent paralyses are rare. After a shorter or longer time, the paralysis of the muscles of the eye disappears. Difference in the size of the pupils is common, reflex rigidity very rare. *Charcot's* cardinal symptom, *nystagmus*, is observed quite frequently. First visible in movements, in which the already paralyzed muscles are used ("nystagmoid twitchings") it becomes more distinct later on and is noticeable even when the eyes are at rest. Nystagmus horizontalis, as well as nystagmus verticalis and rotatorius may be observed; the nystagmus is most frequent when the patient looks to one side. *Redlich* regards it as completely analogous to intention tremor.

Paralytic weakness of one extremity, especially of one leg, has often been observed as an initial symptom. I have observed at the same time three cases with initial paresis of one extremity (twice of a leg, in one case of an arm). In addition to the weakness, *rigidity* of the musculature is usually also present.

The presence of a spastic paralysis in the lower extremities, is quite frequently observed in multiple sclerosis. *Charcot* already pointed to this and later observers have, for the most part, confirmed it. Occasionally this condition is only suggested and recognized with difficulty—in other cases, it is well developed.

Increase of the patellar reflexes and ankle clonus belong to the most frequent and important early symptoms of disseminated sclerosis. The increase in the reflexes may be demonstrated on both sides. In the early stages also tremor in one extremity is not rare.

If the disease is fully developed, the cardinal symptoms, postulated by *Charcot*, are present—*nystagmus*, *intention tremor*, *scanning speech*, to which

are further added *reflex exaggeration* in the lower extremities with *loss of the abdominal reflex.*

The so-called intention tremor is a coarse trembling of the extremities while performing voluntary movements; the trembling becomes more and more pronounced, the nearer the extremity approaches the goal which it is endeavoring to reach. At the same time, muscle groups lying at some distance are contracted. If in a relatively severe case, with the patient in a sitting posture, he attempts a movement with one extremity, his entire body begins to tremble (wave to and fro). If the patient takes hold of an object, it is swung to and fro with the extremity, but usually held fast, in as much as intention tremor is never associated with any considerable weakness of the extremities. The number of the contractions is not very large, much smaller than in Basedow's disease.

The head also takes part in the oscillating movements. In more severe cases, the tremor in the trunk becomes very decided and during intended walking the trembling of the legs is also very distinct in most of the cases. The tremor does not stop, even if the movements are frequently performed in succession, but the intention tremor disappears altogether, when the patient is in repose.

Scanning speech is a very noticeable disturbance. The speech becomes monotonous and its capacity for modulation is absent. It becomes drawn out as each syllable is clearly separated from the one following. Even though this speech disturbance sometimes appears early, in the majority of cases, scanning speech is a symptom, that becomes pronounced only at the height of the disease. *Bradyphasia* is occasionally present without synchronous scanning (*Redlich*). *Dysarthria* is no rare symptom.

The exaggeration of the tendon reflexes has already been mentioned. An important symptom is the diminution of the cutaneous reflexes, especially of the *abdominal reflexes* (*Müeller*), which may appear early.

Besides the increase of the reflexes, especially in the lower extremities, other symptoms as well point to lesions of the pyramidal tracts. Thus the *Babinski phenomenon* is present, and dorsal flexion of the great toe appears not only when the test is made in the typical way (stroking the inner margin of the foot or the instep or above the metatarsus quintus), but sometimes even in walking, if the patient sets his foot down firmly. *Oppenheim's leg phenomenon* is clearly developed (dorsal flexion of the great toe upon vigorous stroking of the edge of the tibia). The testing for Mendel's reflex upon the dorsum of the foot occasionally elicits plantar flexion of the great toe by tapping the dorsum of the foot at the base of the third and fourth metatarsal bones (whereas in healthy people, a dorsal flexion results).

Disturbances in sensation are more frequent in the course of the disease, than was formerly thought. Disturbances in perception, however, are only temporary, and severe only in exceptional cases.

Phenomena appear due to sensory irritation, especially intercostal pains, pains in the joints, paræsthesias, and neuralgic troubles. In one of my cases neuralgia of terrible severity persisted for weeks. *Oppenheim* reports the case of a patient in whom "tic douloureux" was an initial and permanent symptom of the multiple sclerosis.

Sensory phenomena of functional loss usually do not persist for any length of time. They are not very serious and cause only a deadening of sensibility, no complete loss of sensation. The sensory disturbance may be uniform and affect the various qualities of (superficial as well as deep) sensation in the same way, or there may develop a partial disturbance in sensation, which affects only the pain or temperature sense and disappears after a shorter or longer duration (*Freund*). They may then return anew and disappear again. Permanent disturbances of sensation are rare.

Oppenheim observed the Brown-Séquard symptom complex as a transient phenomenon and also hemianæsthesia.

Paræsthesias, like formication, feeling of numbness, are not very rare.

Rectal disturbances are observed at the beginning of the disease only in exceptional cases. In the late stage of the affection incontinentia alvi is frequently seen. I have observed it in quite a large number of cases. The patients, as a rule, perceive the passage of the feces, but can not prevent it. I have often seen this as a transient symptom.

Bladder disturbances are frequent. *v. Frankl-Hochwart* and *Zuckerkandl* found them in four-fifths of their cases, especially when paraplegia of the lower extremities existed. They may be the very first signs of the disease, which is followed by others only after months, or even years. The form of the bladder disturbances is not always the same. At one time simple incontinence with dribbling of the urine appears; at another, spasms of the sphincter with intact sensibility of the bladder, which demands catheterization, or there develops the "irritable bladder" with frequent tenesmus and incomplete emptying and a sudden shifting between incontinence and retention.

The bladder disturbances may disappear permanently or return after a shorter or longer time. Persistence of the bladder disturbances is noted, as a rule, only towards the end of the disease.

Bladder disturbances are more frequent than rectal disturbances according to my observations, and affect many patients in the initial stage of the disease.

Disturbances in the realm of the *sexual sphere* are much less frequent than might be suspected, considering the frequency of the bladder disturbances. Only patients with far advanced disseminated sclerosis complain of frequent pollutions or unsuccessful cohabitation. One of my patients married some years after the beginning of his disease, and begot a goodly number of children.

In some cases, *bulbar disturbances* develop. Sudden paralyses of the soft palate, of the pharyngeal musculature of transitory character are thus explained. Disturbances in deglutition and mastication, regurgitation through the nose, faulty swallowing, may be present as initial symptoms, but do not remain. Disturbances of phonation may be caused by paresis of the laryngeal musculature, and it is especially pareses of the interni, rarely paralyses of the vocal cords, that are observed. Trembling of the vocal cords in phonation has likewise been described. *Oppenheim* describes tremolo of the voice in uttering ā (German E).

Like other motor paralyses in disseminated sclerosis, the bulbar paralyses do not persist; I myself have seen the most severe bulbar processes retrogress. Towards the end of the disease, these phenomena may become permanent and present themselves in a form clinically resembling progressive bulbar paralysis, but without muscular atrophy in the region of distribution of the bulbar nerves.

Obsessional laughing and crying are sometimes observed very early. If they exist, they sometimes persist permanently. One of my patients was first attacked by obsessional laughing at a funeral, as he stepped up to the open grave, and has now suffered for ten years, uninterruptedly, from this very annoying symptom. The transition from compulsory laughing to compulsory crying may take place very suddenly.

Apoplectiform attacks, which threaten the patient in every stage of the disease, but luckily, too, spare many patients, play an important part. The attacks are occasionally accompanied by complete loss of consciousness, and are then accompanied in some cases by epileptiform seizures. Or, it may be that only severe attacks of vertigo occur, which may be compared to apoplectiform attacks, strike the patient suddenly, and if he is standing or walking at the time of the seizure throw him to the ground. In direct connection with these attacks, one sees more or less extended unilateral paralyses of the cerebral nerves. Or immediately after the attacks, combinations of cranial-nerve palsies will be observed, which show extraordinary variations. All the cerebral nerves, even the auditory, are affected occasionally. Hemiataxia after an attack has been described by *Oppenheim*, who reports, too, rarer, suddenly occurring paralyses (atrophic paralysis with ataxia of one arm). I have seen the clinical picture of an acute extended bulbar paralysis with paralysis of deglutition, of the soft palate and of the larynx.

Ataxia is occasionally present. In such instances it affects either only single extremities, or one-half of the body, or finally, only the lower extremities. It is of the nature of cerebellar ataxia (*Redlich*), and manifests itself by a tottering gait. According to *Oppenheim*, ataxia must be assumed to be present if the disturbance increases when the eyes are closed. Ataxia does not by any means exclude the existence of spastic conditions in the

limbs. Its acute development in the upper limbs with or without bulbar symptoms has been observed several times.

Disturbances of the intellect, especially failing memory, are common; delirium, and dementia are present only in exceptional cases; if either occurred, they were, as a rule, in most of my cases, transitory phenomena.

Complaints about *headaches* are common and they occasionally resemble migraine.

Respiratory disturbances are unusual. Towards the end of life Cheyne-Stokes respiration is observed. *Oppenheim* has described conditions of asphyxia.

The *pulse rate* is accelerated, if a bulbar lesion is present; tachycardia is often continuous, but remains within moderate bounds.

Muscular atrophies are very rare. Should they develop, however, the electrical excitability is changed only in exceptional cases. Atrophy is seen in individual groups of muscles, or possibly in the musculature of an entire limb.

The paresis usually affects both legs, so that spastic spinal paralysis results. As a matter of fact, in many spastic spinal paralyses, the presence of a disseminated sclerosis is only established postmortem (as long as these peculiarities were unknown). Unilateral forms with paralytic phenomena and tremor in one-half of the body were observed by *Bikeles*, *Oppenheim*, *Edwards*. Extensive atrophic pareses, concerning which *Probst* reports, are very rare. In his case, the symptom complex of amyotrophic lateral sclerosis was present.

Course.—With respect to the course, three forms may be distinguished: 1. Forms with an insidious onset. 2. Cases with acute initial symptoms, but slow course with remissions and exacerbations. Both forms appear preponderantly in young persons. 3. Acutely beginning and quickly ending cases ("acute multiple sclerosis").

The cases with an insidious onset seem to be the more frequent. Since the symptoms develop very gradually and show a considerable tendency to remissions, the disease in most cases is supposed to be functional. This error is supported by the fact that the disease is nearly always combined with hysteria.

In the discussion of the individual symptoms, the possibility of an acute onset was mentioned several times. With this onset the initial picture may be very varied and paralyses of the most varied kinds may develop (unilateral paralysis of the body, ataxia of single limbs, bladder and rectal disturbances, sensory paralyses and those of the muscles of the eye, bulbar paralyses, etc.).

Extraordinarily important for the recognition of the nature of disseminated sclerosis is the peculiarity of the disease, in having long remissions and in advancing intermittently.

The remissions may be surprising even in advanced cases, and in less progressive ones create the impression of a cure.

Exacerbations follow especially under the influence of weakening diseases, of the puerperium, great emotional excitement and physical over-exertion. Especially injurious seems to be the accumulated emotional excitement of the newly married. Within a short time, I have seen serious symptoms appear in six young brides whose anamnesis proved that the beginning of the disease dated far back into their girlhood.

With remissions and exacerbations the disease gradually grows worse. The duration is quite long, amounting to 5 to 10 years, and even longer.

Death results from intercurrent affections, or because of the bulbar changes, occasionally during an apoplectic attack.

One form of the disease terminates in death very quickly, at most within a few months, or a year: then one speaks of *acute multiple sclerosis*. *Marburg, Schlagenhaufer, Gudden, Flatau* and others have especially studied this form. The development of some of these cases from an inflammatory process, encephalomyelitis, is proved with considerable certainty by several observers. I myself have observed such a case clinically and anatomically. *Oppenheim* has repeatedly seen the development of a disseminated sclerosis from an infectious myelo-encephalitis.

Differential Diagnosis.—Many affections are to be considered in making a differential diagnosis. Of especial importance is the differentiation from hysteria. For a number of years, I have been emphasizing in my lectures, again and again, the frequency of these confusions; *Redlich and Oppenheim* have had the same experience. In every case of hysteria with ankle-clonus the possibility of a disseminated sclerosis must be considered; furthermore, also in every case of hysteric amaurosis, in quickly disappearing disturbances in gait and pareses in hysterical individuals, should signs of an affection of the pyramidal tracts be present. If the *Babinski* phenomenon is positive with dorsal flexion of the great toe, likewise *Oppenheim's* leg phenomenon, if there be spastic paresis in the lower extremities, an organic affection must be assumed in addition to hysteria and then multiple sclerosis stands in the front rank of probability. This probability is increased if the patient attacked is a young girl or a young woman. The further course, in particular transient bladder disturbances, enables the physician to make a diagnosis.

Differentiation from *encephalitis* may become very difficult, especially if the symptoms of an acute pontile or medullary affection appear in young persons without affection of the vessels or heart and there is no kidney disease or syphilis. Detection of an elevation of temperature from a directly preceding infectious disease would speak rather in behalf of the diagnosis of an encephalo-myelitis.

If in a young person, the acute lesion (in the form of an apoplectic attack)

affects the brain, the further course of the disease must be awaited if the other changes discussed above are absent. If the symptoms disappear, and absence of fever and a directly preceding ætiological factor is noted, a progressive paralysis or multiple sclerosis is probable. If the intellect is not seriously disturbed by the attack, disseminated sclerosis is the more probable diagnosis.

But if a lesion in the vessels (atheroma) is present, even with multiple foci, the decision must be in favor of assuming the presence of a softening, since multiple sclerosis is more apt to be found in young people.

Cerebro-spinal syphilis can give rise to a disease picture very similar to that of multiple sclerosis. Besides the anamnesis, the effect of specific treatment will be of differential diagnostic value (though such treatment must be used with caution).

Cases of *solitary* as well as *multiple tumors of the brain* have been observed, which clinically bore a close resemblance to disseminated sclerosis (*Bruns*, *Nonne*); in the latter even choked disc has been observed. Of course this is not a common occurrence. The absence of disturbed intellect and of constant headache militates against a diagnosis of a tumor, whereas signs of constantly increasing intra-cranial pressure make the existence of a tumor more probable.

Many years ago, *Charcot* separated multiple sclerosis and *paralysis agitans* diagnostically in their clinical phenomena. Paralysis agitans is a disease occurring in older persons; the tremor of shaking palsy is unlike intention tremor; nystagmus and scanning speech do not belong to paralysis agitans, while the general rigidity is not peculiar to multiple sclerosis.

Friedreich's disease, or really *heredo-ataxie cerebelleuse* (*Marie*) which is distinguished from it by many authors, may closely resemble certain stages of multiple sclerosis. In heredo-ataxia there is nystagmus and a disturbance in speech, which recalls scanning speech; the tendon reflexes may be increased. A choreiform tremor can affect almost the entire musculature of the body and become intensified upon motion. The gait, however, is ataxic, occasionally a peculiar club foot develops and an optic atrophy, which does not improve.

Progressive paralysis quite often exhibits a certain similarity to multiple sclerosis. But the stumbling over syllables, the reflex pupillary rigidity, the trembling speech, early psychic disturbances, unequal innervation of the facial musculature, and the trembling of the lips are peculiar characteristics of the paralysis and not of multiple sclerosis.

Differentiation from the affection, which was described by *Westphal* as *pseudo-sclerosis* and later was specially studied by *Strümpell* and *v. Frankl-Hochwart*, may prove difficult. These are comparatively rare cases, presenting many of the clinical symptoms of disseminated sclerosis, while no

anatomic change can be found postmortem. This affection is distinguished from multiple sclerosis by the absence of nystagmus, of the optic affection and also of the genuine spastic phenomena. The early development and the severe psychic disturbances (dementia) are conspicuous; *Strümpell* emphasized, besides, the fact that the tremor appeared even during rest, was slower, but of greater amplitude. The active movements also are decidedly retarded. Speech is scanning, but also clearly dysarthric. Epileptiform and apoplectiform attacks are present in almost every case. On the basis of these phenomena, correct diagnoses were made in vita by *Strümpell* and *v. Frankl-Hochwart.*

At present, *pseudo-sclerosis* cannot be distinguished from genuine diffuse cerebral sclerosis with degeneration of the pyramidal tracts (*Strümpell, Oppenheim*). The latter author is also of the opinion that the combination of progressive spastic paralysis with progressive dementia in childhood must arouse a suspicion of diffuse cerebral sclerosis.

Therapy.—One is, alas, comparatively helpless, face to face with the development of the disease, and can only sometimes, guided by a knowledge of the peculiarities of its course, obtain remissions.

I have seen but few benefits follow the use of medicines. With anæmic persons, especially in cases of a low state of nutrition, preparations of iron and arsenic may well be used. In such cases I prefer ferratin, ferrum cacodylicum or ferrum lacticum in combination with preparations of quinine, or prescribe Blaud's pills; arsenic I prefer to administer subcutaneously, intramuscularly, as sodium cacodylate (every other day 0.02 in a watery solution), or as atoxyl (every other day 0.05–0.10 in a watery solution). Oppenheim has seen bad results from mercurial treatment.

Of great importance is the carrying out of a strict rest cure. In poorly nourished patients synchronous forced feeding is of considerable value. The use of (not prolonged) lukewarm baths or baths of carbonic acid (lasting from 10 to 15 minutes, temperature between 88° and 94° F.) seems to me advantageous since they often influence the rigidity very favorably. If the treatment by forced feeding and rest is continued long enough, and the patient takes very good care of himself afterwards, a long remission may appear. At present I am treating a young woman, who was paraplegic four years ago, had bladder trouble and had so far recovered by means of forced feeding and rest, that she could manage the household for years, until over-exertion and excitement paved the way for a return of the paraplegia. Such long remissions I have repeatedly seen.

I have seen strikingly favorable results from treatment by baths in Bad Gastein.

Massage seems to act favorably. *Oppenheim* recommends that passive movements be used in the bath.

Steam baths, hot baths, affect unfavorably the course of the disease;

deterioration may follow immediately upon the bath. The same holds true of all exciting hydriatic and other procedures.

Physical over-exertion may induce rapid deterioration; likewise undernutrition and psychic excitement may cause rapid progress of the disease.

Strong spices, highly seasoned food and alcoholic drinks are to be avoided.

Sexual excesses in one of my cases, caused in a short time complete development of the disease which had been up to then but slightly troublesome.

The abuse of tobacco also seems to be followed by an advance in the disease.

4. SYRINGOMYELIA, SPINAL GLIOSIS

BY

HERMANN SCHLESINGER (Vienna)

Pathological conditions of different ætiology are embraced by the term syringomyelia. They lead to the formation of long, extensive cavities, which with special frequency occupy the central sections of the spinal cord. They also cause considerable tumor-like proliferations of the glia. We are dealing altogether with progressive processes; the stationary or retrogressive cystic degenerations of the spinal cord are to be separated from syringomyelia (*Kienböck*). The clinical picture is very variable, but permits common traits to be recognized in the individual cases.

Pathological Anatomy and Pathogenesis

The spinal cord is frequently flattened out for long stretches, and much reduced in substance; it is true, it frequently appears swollen in smaller sections. If the cavity extends to the medulla oblongata, the latter becomes very asymmetrical. Only in a minority of cases does one find a diffuse tumor-like swelling of the spinal cord with cavity formation. In cross-section, one sees a cavity occupying the central portion or even a large part of the dorsal portion of the spinal cord; the size of the cavity varies greatly at levels but little distant from each other. Frequently there are no large cavities at all and instead one finds a long extended tumor, which occupies the central area of the spinal cord, appearing now like a hard narrow glia tube, now presenting a decidedly tumorous appearance. Pure unilateral localization of the disease is rare; even then the gray matter is preferably attacked (posterior horn). Most frequently, the cervical portion of the cord, the cervical enlargement and the upper dorsal segment are the seat of the lesion. Often, the extent, lengthwise, is very considerable and the changes go far down into the lumbar segment, indeed even into the sacral portion at one end and into the bulbus medullæ at the other. Cases in which the dis-

ease is limited to the lowest sections of the spinal cord or to the medulla oblongata alone, are of rare occurrence.

Most frequently injured are—the region of the central canal, the posterior horns and the posterior columns; the latter are sometimes, for long stretches, entirely swallowed up in the formation of the tumor or cavity, but usually suffer extensive destruction only in their ventral parts (Fig. 76). The lateral pyramidal tracts are, as a rule, interrupted at several points, and show descending degeneration on one or both sides. The other parts of the cross-section, as the lateral cerebellar tracts and the anterior columns, are affected less frequently, but at any rate occasionally, while the anterior horns are affected quite frequently.

In the medulla oblongata, the lesion very rarely (*Spiller* describes such a case) extends beyond the lower margin of the pons in the direction of the

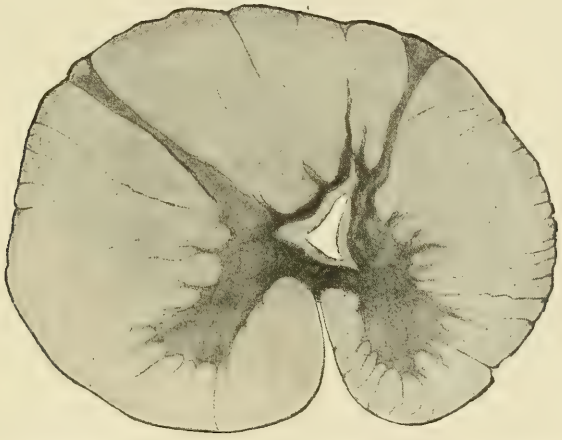


FIG. 76.—Syringomyelia. Cavity behind the central canal. (*After Schmaus.*)

cerebrum. The changes occupy, preferably, definite places of the cross-section (*H. Schlesinger*); usually either lateral or medial fissures are present. As a rule, the lemniscus, the nuclei of the vagus, the accessorius, glosso-pharyngeus, hypoglossus are affected, often, also, the spinal root of the fifth nerve is destroyed (Fig. 77) (*Schlesinger, Hatschek, A. Westphal, Philipp-Oberthür*).

The tumor is built up of glia cells and glia fibres; likewise the connective-tissue elements play an important rôle. The cavities frequently have an ependymal lining for a shorter or longer part of their course.

The origin of syringomyelias through the breaking down of these tumor-like glioses was pointed out first by *Simon, Westphal*, later by *Schultze, Hoffmann, Oppenheim, Schlesinger and others.*

The condition is termed a *hydromyelia*, if the cavity is lined throughout with ependyma, a *syringomyelic gliosis* (*Schlesinger*), if the cavities are combined with gliosis or have originated in the latter. The fissures of the

spinal cord are surrounded by indurated masses of glia connective tissue, while the syringobulbia is, as a rule, bounded by degenerating nerve tissue. In the cavities and also in their walls are often found residues of hemorrhages and changes in the vessels. The glioses do not extend to the meninges. If extensive meningeal changes with cavity formations do exist, the condition is not one of glio-syringomyelia.

As predisposing to the development of syringomyelia, we might consider anomalies in the development of the entire central nervous system or of the central canal (*Leyden*) and its immediate vicinity. Most likely glioses (tumor-like, degenerating proliferations of glia) come from a congenital or early acquired disposition (*Hoffmann, H. Schlesinger*). Traumatism seems occasionally to lead to the development of glioses (*Minor, Westphal*), but a certain predisposition is probably essential.

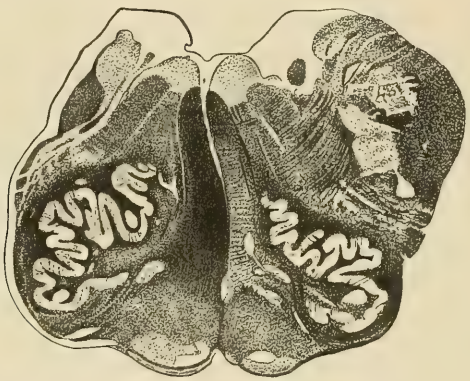


FIG. 77.—Syringobulbia with degeneration of the lemniscus. (*After Schmaus.*)

F. Schultze has made it appear probable that traumatisms during birth form the starting point of the affection. In compression of the spinal cord, a syringomyelia is found occasionally above the place of compression. Infectious diseases bring a gliosis in their train relatively often. Syphilis (*Schwarz, Simon*) and meningitis may be followed by syringomyelia; possibly meningeal and spinal affections are co-ordinated (*Müller, Meder and others*). Changes in the vessels can also, especially in advanced age, cause formation of fissures (without gliosis). The ætiological rôle of inflammatory spinal processes, for instance, myelitis (*Joffroy, Hallopeau*), for the origin of syringomyelia is still debatable.

For syringobulbia, changes in the vessels are of importance, as well as anomalies in development.

The anatomic ætiology of syringomyelia is therefore not uniform.

Besides infections, intoxications might also be an ætiological factor here and there. But this influence is occasionally exaggerated. For the production of a syringomyelia by peripheral traumata there is, as yet, no proof.

Symptoms.—The clinical picture of syringomyelia is extraordinarily varied. The phenomena that come under observation belong preponderantly to the following groups of symptoms: 1. Phenomena in the domain of the motor nerves; 2. disturbances in sensation; 3. vasomotor trophic disturbances; 4. bulbar symptoms.

The motor disturbances are mainly muscular atrophies in the upper and frequently, also, spastic conditions in the lower extremities.

The muscular atrophy begins most frequently insidiously in the musculature of the hand. No other spinal affection leads so frequently in young persons to atrophy of the smaller muscles of the hand. Since the muscles do not always become atrophied in the same order, various positions of the hand can result. The interosseous spaces frequently sink in, the fingers

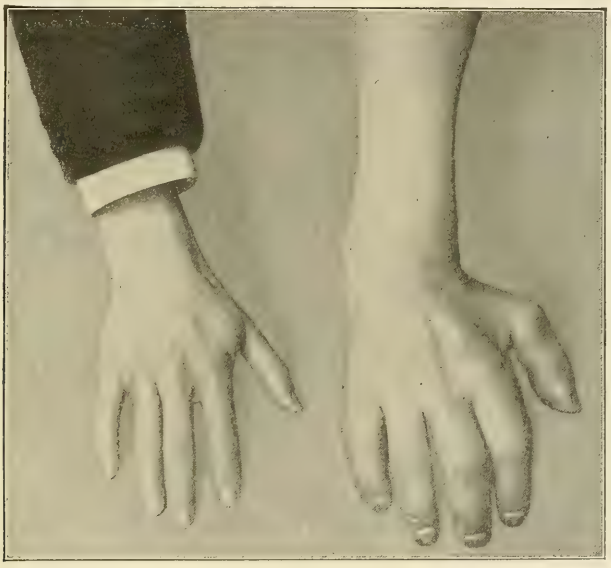


FIG. 78.—Cheiromegaly in syringomyelia (as compared with the normal hand).

are over-extended at the metacarpo-phalangeal joints, flexed at the interphalangeal joints, the tendons of the flexors become prominent in the hollow of the hand, thenar and antithenar muscles become atrophied, so that the position known as "claw-hand" results, or through atrophy of the ball of the thumb and the antithenar eminence, with a change in the position of the thumb, there develops the "ape's hand." Or through early paralysis of the flexors of the wrist-joint, the dorsal musculature of the forearm becomes the stronger, thus causing the development of the so-called "preacher's hand."

In many cases, however, the muscular atrophy begins in the muscles about the shoulder-girdle and extends to the musculature of the arm or of the hand only after a longer time. Scapular "angel's wings" are very

frequent in these cases since the muscles, that hold the scapula in its position, become atrophied. The cucullaris is frequently atrophied only in its lower portion.

It is much less common for atrophy to appear early in the lower extremities; it can then lead to the formation of a pes equinus. The atrophies are mostly asymmetrical.

In the atrophic muscles fibrillary twitching is commonly observed. Reactions of degeneration cannot always be demonstrated.

Rigidity is often present in one or both of the lower extremities. The rigidity can spread to the larger part of the *musculature* of the body, so that the patients find themselves unable to move (*Raymond, Déjérine, Redlich*).

The tendon reflexes in the lower extremities are increased. Ankle clonus is regularly present. In the areas of the muscular atrophies, the tendon reflexes are usually diminished.

Oppenheim's leg phenomenon and the *Babinski* toe reflex (dorsal flexion of the great toe) are present.

Spontaneous movements are observable in all stages of the disease, especially in the fingers and thumbs (*Schwarz*).

In muscles, not wholly able to perform their functions, there occasionally appear symptoms peculiar to myotonia, a condition that I designated as myotonia syringomyelica (observed by *Rybalkin, Fuchs* and others).

The *disturbance in sensibility* is only partial in the great majority of cases. As a rule, analgesia and thermo-anæsthesia are present, while the other sensations suffer qualitatively none or but slight disturbances ("syringomyelic dissociation of sensation," first established by *Kahler and F. Schultze*). The partial paralysis of sensation is demonstrable over a larger or smaller area of the surface of the body. It shows as a rule (*Laehr, Hahn, Déjérine, v. Sölder, H. Schlesinger and others*), segmental arrangement and also an extension corresponding to the segments of the spinal cord. The area of its extension does not necessarily correspond with that of the localization of the muscular atrophies. The disturbances in sensibility may also affect the pain sense alone, even the sensation for cold and heat alone.

The defects in sensibility are also asymmetrical; a sensory hemiplegia has been repeatedly observed.

The thermo-anæsthesia leads to the burns that are so frequent in syringomyelia; they occur in typical places (hands, fingers, shoulder blade, buttocks).

Tactile sensibility and the muscular sense are not infrequently disturbed, but the syringomyelic dissociation dominates.

In the initial stage, sensory irritation phenomena (paræsthesias affecting the temperature sense) are usually present.

The sensibility of the bones is often considerably lowered (*Egger, Neutra*).

The trophic disturbances affect the skin, bones and joints.

The affections of the skin are so manifold, that practically the entire subject matter of dermatology would have to be discussed, if one wished to enter upon this phase.

The urticaria-like eruptions (dermographism), the formation of bullæ which always return in the same places on the body, are of especial importance. Decided thickenings (up to one centimeter) in the skin of the palm of the hand approach to the forms of Morvan's type. Deep rhagades with but slight tendency to heal, complicate this change. Vitiligo, sclerodermic changes, discolorations of the skin, especially in the hands, are common. Peculiar is the "main succulente" described by *Marinesco*, in which through a transformation of the tissue of the subcutis, the skin has a pasty appearance, the atrophies being masked. But there is actually no edema of the skin. The change takes place also in the skin of the fingers. Very frequently there develop in the fingers *painless* whitlows, which, if repeated, cause extraordinary malformations of the fingers and the hand. Not rarely, in these painless inflammatory processes, phalanges are lost. The nails become misshapen, claw-like, rudimentary. Perforating ulcer of the sole of the foot is rarer and must always remind one of a process which clinically closely resembles syringomyelia—namely, leprosy.

Extensive spontaneous gangrenes occur, but are not peculiar to syringomyelia.

The disturbances in perspiration are very interesting. The secretion of sweat is increased, or anidrosis develops. I also have described "paradoxic sweat secretion," perspiring under the influence of cold. The anomalies of perspiration affect only individual sections of the surface of the body, the demarcation of which is parallel to the spread of segmental disturbances in sensibility.

The most striking *affections of the bones* are spontaneous fractures, which occur preferably in the lower extremities; their whole course is unattended by pain and they result mostly from slight traumatism. Union often takes place slowly with formation of a large amount of callus. The bones of the forearm are preferred.

The fracture is remarkably often a pure transverse fracture (*Gnesda*). *Tedesko* found in the systematic X-ray testing for transparency in the bones of syringomyelic patients many cases with changed structure.

Spontaneous bone necroses of varying extent have been described several times, likewise formation of multiple exostoses.

Scoliosis is often present, possibly in a quarter of the cases (*Bernhardt*); it can attain high degrees, is combined with kyphosis and predominant in the thoracic spinal column. The kypho-scoliosis usually develops slowly, but may be an early symptom (*Charcot, Oppenheim, Roth*).

The chest is often deformed, a trough-like sinking in at the upper sternal

end and the neighboring ribs ("Thorax en bateau"—*Marie, Astie*) is considered characteristic for syringomyelia.

In the course of the disease, a hand occasionally grows to gigantic size, but not regularly; cheiromegaly (*Hofmann, Marie*) (Fig. 78). The partial gigantic growth may also affect one foot—podomegaly (*H. Schlesinger*). The typical picture of acromegaly is, however, not simulated by these conditions.

Bladder and rectal disturbances belong to the early symptoms only in the lumbo-sacral forms of syringomyelia; otherwise they develop, if at all, only in the later stages of the disease. Sphincteric spasm, afterwards incontinentia urinæ are the most frequent anomalies. The cystitis may run a wholly painless course.

Diabetes insipidus, glycosuria, were observed in a number of cases. I have seen nephrolithiasis in several cases.

Disturbances in the realm of the sexual sphere are unusual.

Hemiatrophia faciei has been observed as a striking complication in several cases.

Bulbar lesions are common clinically as well as anatomically. They are either unilateral or asymmetric.

The *tongue* is often affected. Then there develops a hemiatrophy with wrinkling of the mucous membrane, fibrillary twitchings and considerable diminution in size of one-half of the tongue (*Lamacq, Maixner, Hitzig, Raymond, Schlesinger*).

Hemiatrophia linguæ is often combined with unilateral paralysis of the musculature of the palate and the larynx, so that the symptom-complex of unilateral bulbar paralysis is present.

The disturbances in deglutition are occasionally considerable: faulty swallowing, regurgitation of liquids through the nose, remaining of bits of food upon the base of the tongue. They may be transitory.

The laryngeal paralyses are characterized by a total palsy of the recurrent laryngeal nerve with simultaneous paresis of the soft palate and of the muscles of deglutition (*H. Schlesinger, Baurovicz*). Atrophy of a vocal cord is common; the external branch of the recurrens need not be injured.

Paralyses in taste occur, sometimes in individual qualities of taste.

Speech may be decidedly bulbar or hoarse with slight change of voice (consequence of the recurrens paralysis).

Often the bulbar symptoms appear with an intense *attack* of *vertigo* without loss of consciousness. The feeling of dizziness is of extraordinary violence, so that the patients fall to the ground. Immediately after this feeling of vertigo, the bulbar paralysis appears, or the symptoms of it already present become more progressive.

In many cases, a unilateral lesion of the *sensory trigeminus* is the first symptom of bulbar disease (the spinal trigeminal root is attacked). The

extension of the disturbances in sensation in the scalp, and in the face follows with sharp boundaries that are not to be found in peripheral trigeminus lesion. The disturbance in sensation, which in most of the cases is partial, is bounded, as it advances, by lines which are packed closely into each other, in the form of an onion, about the mouth or the nose, and which have

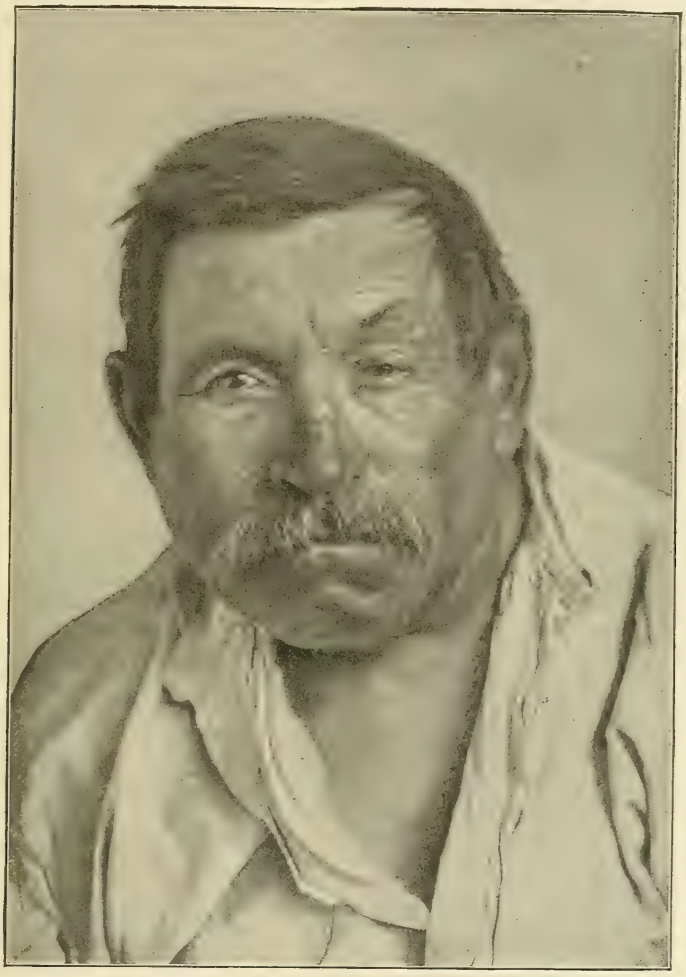


FIG. 79.—Paralysis of the sympathetic at the left side in a case of unilaterally localized syringomyelia (gliosis unilateralis). (*After Heinr. Curschmann.*)

been closely studied by *v. Sölder* and myself. This type of disturbance in sensibility corresponds to the segmental arrangement of the elemental constituents of the trigeminus.

First, in regular order, the laterally situated parts of the skin of the face, and only towards the end, the most medially situated sections become anæsthetic, or only analgetic and thermo-anæsthetic.

Neuralgia-like pains in the face are present, especially in the beginning of the bulbar affection.

Trophic disturbances (corneal affections) which often accompany a trigeminus lesion, are rare.

Facial paralyses are rarer, may strike now only the buccal branch, now the entire facialis. The paralysis is located on the side of the more pronounced defects in sensibility in the trunk.

The *ocular disturbances* are of various kinds. Very common is nystagmus or nystagmus-like twitchings in turning the eyes to either side. Of paralysis of the muscles of the eye, the abducens paralyses are most frequent; the *Argyll-Robertson* phenomenon, which has been observed several times, is, as a rule, to be traced back to a complicating disease (tabes or progressive paralysis).

Very frequently, in about one-sixth of the cases, a *paralysis* of the *sympathetic* with all its typical phenomena (sunken eyeball, moderate degree of ptosis, contraction of pupils, which retain the ability to react) is present (Fig. 79). Besides, there are vasomotor phenomena in the same half of the face and unilateral disturbances in the secretion of sweat (anidrosis or hyperidrosis).

The visual field, in the great majority of cases, where there is no complication with hysteria, is normal. Neuritis and choked disc have been described several times.

The bulbar disturbances, therefore, affect mainly the fifth to the twelfth cerebral nerves.

Common Clinical Types.—Very frequently one sees a syringomyelia with the "classic symptoms" when the disease is confined to the cervical segment of the cord. Then there are present, usually, muscular atrophy in the hands of the Aran-Duchenne type, dissociated disturbances in sensibility and trophic-secretory disturbances in the upper half of the trunk and the upper extremities. Or the muscular atrophy begins in the musculature of the shoulder-girdle and is accompanied in earlier or later stages by disturbances in sensation (*humero-scapula type*).

Often in the early stages of the disease, the disturbances in sensibility are but slightly developed and their existence demonstrated with difficulty.

Symptoms of spastic spinal paralysis, or of an amyotrophic lateral sclerosis are not very rare. In both cases disturbances in sensation may long be absent or be only slightly developed, and the diagnosis will have to be made from trophic disturbances of the skin, the bones, or the joints.

In the lower extremities also, onset with the well-known triad of symptoms is possible. Bladder disturbances are more common in these forms than in others, just as is perforating ulcer of the foot. The syringomyelic pes varo-equinus is usually found with this form (*lumbo-sacral* syringomyelia).

I have already mentioned the syringomyelia, that is confined to the

lowest segments of the spinal cord (*sacral form*). In one case observed by me, there were constantly present recurring perforating ulcers in both heels, disturbances in the sexual sphere, loss of libido and ability of erection,

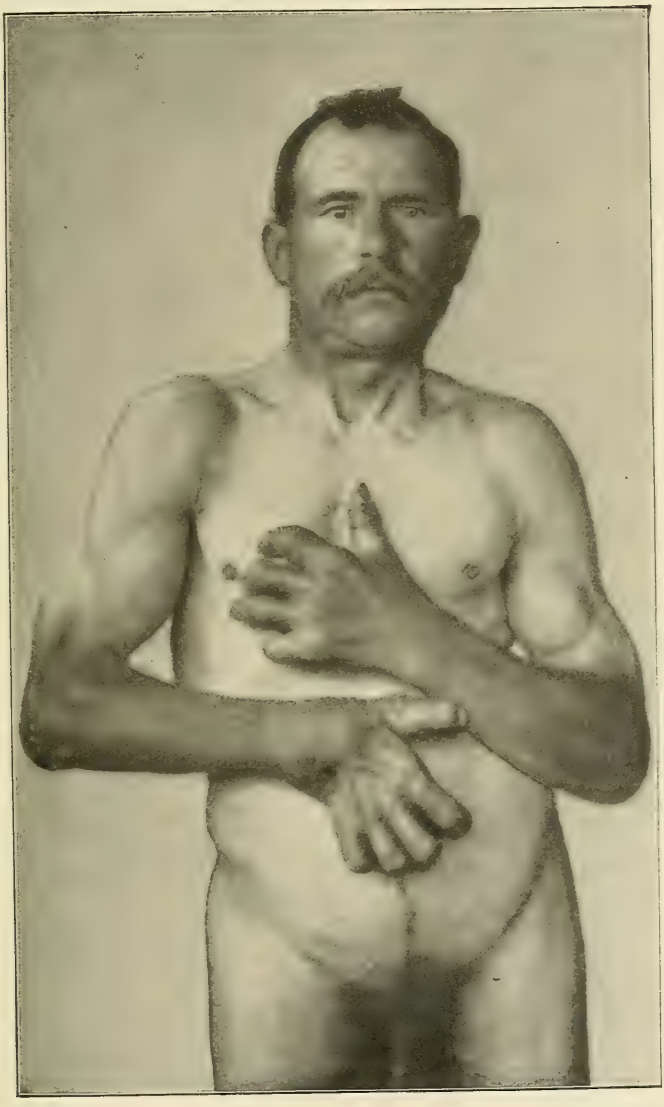


FIG. 80.—Syringomyelia of the Morvan type. (*After Heinr. Curschmann.*)

partial paralysis of sensation in the perineum and about the anus, loss of the feeling of a full bladder and loss of the Achilles tendon reflex.

Beginning of the disease in the medulla oblongata, and a later affection of the medulla spinalis is far less frequent than secondary syringobulbia.

Sometimes the gliosis attacks a posterior horn to a considerable extent

lengthwise (*Rossolimo, Oppenheim*); then there occur unilateral disturbances in sensibility, which remind one very much of hysteric disturbance. Only in rare isolated cases has a general anæsthesia been observed in syringomyelia.

Through the predominance of trophic phenomena, peculiar clinical forms are created, as the *osteo-arthritic* form due to early and extensive changes in bones and joints. Cases with serious malformations of the fingers and hands, as a result of repeated whitlows and phlegmonous processes are particularly frequently observed. These forms with unsightly thickenings and distortions of distal parts of the extremities and severe disturbance of tactile sensibility are designated as *Morvan's symptom complex* (Fig. 80).

The *tabetic type* may originate from a combination of tabes and syringomyelia (for instance tabetic phenomena in the legs, syringomyelic in the arms) or by localization of a gliosis in the posterior columns.

Besides these types, there are rarer forms, caused by peculiar arrangements of the gliosis or by combination with other diseases, as gliosis cruciata (*Oppenheim*) with typical disturbances in the upper and lower limbs of opposite sides. The *pachymeningitic forms* soon lead to the phenomena of transverse section of the cord with motor and sensory paralysis of the lower half of the body.

Combination with hydrocephalus is not unusual and has been observed several times in association with disturbances in the development of the bones and the soft parts.

Oppenheim and Marburg have repeatedly seen "cervical ribs" combined with syringomyelia. Syringomyelia has repeatedly been observed (I have seen it) with cerebral tumors.

The very common combination with hysteria is clinically important, as it modifies many of the phenomena of syringomyelia.

Onset, Age, Course.—The disease, as a rule, begins stealthily, exhibiting a preference for young persons. I have several times been able to recognize it clinically in young children. An onset in very old age or even in a moderately advanced age is less frequent, though the presence of the disease even in old age is not rare. It is usual, however, to find that the phenomena began a long time before. The disease not infrequently lasts from 20 to 40 years. If the disturbances are not serious, it is usually the injuries that drive the patient to the physician. That is why the surgeon is so frequently the first to see the patient. It is comprehensible that some vocations lead to an earlier discovery of the disease (thus cooks and stokers seek a doctor because of their frequent burns).

Syringomyelia is found everywhere, provided that its variable clinical picture is known. In my opinion it stands third or possibly fourth among all the diseases of the spinal cord with reference to the frequency of its occurrence.

The affection has, it is true, a general progressive character, but long

stationary periods, lasting even for many years are seen in many cases. Very long remissions have been repeatedly described (*Hatschek, Bruce, and others*). Acute exacerbations are frequent, being observed especially with localization of the disease in the medulla oblongata and may possibly be the result of hemorrhages, softenings or edema.

Complications may cause death. Among them cystitis with ascending inflammation is specially important, as well as purulent infections of the subcutaneous connective tissue and of the joints. I have frequently seen nephrolithiasis with syringomyelia.

In spite of the frequency of syringobulbia, the fatal outcome is but very rarely caused by bulbar disturbances. Intercurrent diseases, especially tuberculosis and typhoid, add a large contingent to the number of fatal cases.

Differential diagnosis must take into consideration an extraordinarily large number of diseases. Among these the most important practically is leprosy. A very large number of monographs are concerned with this theme, which is of immense importance for the question of the spread of leprosy in civilized countries.

Among the most important we may mention those of *Laehr, Zambaco, F. Schultze, v. Düring, Lie, Bergmann, Jeanselme and H. Schlesinger.*

The diagnostic difficulties are increased by the fact that the combination of leprosy with syringomyelia (*Gerber-Matzenauer*) has been observed with certainty. It is true that such a connection was formerly assumed several times, even postulated as causal (*Zambaco, Marestang*), but this assertion has not been proven. It was supported by the clinical similarity which especially Morvan's type of syringomyelia exhibits to a form of leprosy, but in leprosy, a spinal gliosis is absent from the anatomic phenomena and in syringomyelia, the bacilli of leprosy are absent.

According to the present status of the question, the following factors are to be considered: the origin is now less decisive since new leprous foci are continually being discovered; at any rate, a case originating from a region which has always been free of leprosy, speaks against this disease. Nerve thickenings, especially the spindle-shaped, are peculiar to leprosy; in excised pieces the presence of leprosy bacilli may occasionally be demonstrated. They are early and regularly found in the N. auricularis magnus. Oval cicatricial patches of the skin devoid of pigment with anæsthesia, which have come from pemphigus bullæ, point to leprosy. Tubercular affections of the skin, which are spread over a large part of the surface of the body and also attack the face, are to be similarly interpreted, likewise attacks of fever, especially during progression of such eruptions. A bacteriological examination of the nasal secretions may often early reveal the specific bacillus in leprosy (*G. Sticker*). Peripheral facial paralysis is rare in syringomyelia, frequent in leprosy, especially with marked participation of the frontal branch of the facial. Even specific disturbances of the eyes are not

exactly rare in the latter affection. For gliosis the following speak: bladder and rectal disturbances, increase of the tendon reflexes, spastic phenomena in the lower extremities, severe multiple painless joint affections, the humeroscapular type of muscular atrophy, nystagmus, attacks of vertigo, bulbar symptoms, disturbances in perspiration, segmental disturbances in sensibility, whereas the sensory disturbances in leprosy occur more in discrete areas.

Since syringomyelia may present the symptoms of scleroderma as well as of *Raynaud's* disease and also of pemphigus, differentiation will occasionally be difficult, but can usually be decided on the basis of concomitant symptoms, since gliosis beside these symptoms, exhibits others more peculiar to itself (for instance spastic symptoms, partial paralysis of sensation, bulbar symptoms, etc.), in short, symptoms that do not appear in skin affections.

Arthritis deformans is distinguished from atypical forms of central gliosis by the absence of analgesia, and partial macrosomia by the absence of disturbances in sensibility.

Hysteria is occasionally very difficult to distinguish from syringomyelia, especially from the forms with disturbances in sensation without muscular atrophy. The bulbar symptoms, the increase in the tendon reflexes, sympathetic paralysis, trophic disturbances will, as a rule, make diagnosis easy, though hysteria is often combined with syringomyelia.

Root and plexus neuritis is distinguished by absence of the cord phenomena (reflexes in the lower extremities unchanged), by the violent irritation phenomena, and the uniform disturbance of all qualities of sensation.

Polyneuritis only rarely causes confusion, since its onset is much more quick and it is usually bilateral from the beginning.

A *spondylitis tuberculosa* can more easily simulate the picture of a syringomyelia. The muscular atrophies may be similar, also the dissociation of sensation may be analogous to that in syringomyelia; even the oculo-pupillary symptoms (sympathetic paralysis) occur. But the affection of the bones is recognizable as such by direct inspection or palpation or in the radiograph; the spinal column is held rigidly, transverse section symptoms caused by the cord disease develop rather quickly.

The separation from *hæmatomyelia* will not be very difficult with a knowledge of the history of the case (acute onset, especially after traumatism). Furthermore the phenomena in hæmatomyelia have a tendency to become less, in syringomyelia usually not.

Syringomyelia may be confused with *tabes*, since occasionally the clinical picture of syringomyelia reminds one strongly of that of tabes. Tabetic disturbances of the cerebral nerves (bilateral posticus paralysis, Argyll-Robertson's phenomenon), loss of the tenderness to pressure in the ulnaris, paralysis of the muscles of the eye, and especially oculomotor or bilateral abducens paralyses will suggest tabes first, if the other symptoms are specially

pronounced in the lower extremities. But if a partial paralysis of sensation is permanently present in the lower limbs, if muscular atrophies exist, if the disturbances in the muscular sense are not of a high degree, one will have to think of syringomyelia, even if there be ataxia and loss of the patellar reflexes. Typical phenomena in the upper extremities will confirm with certainty the diagnosis.

From the *spinal amyotrophies* (poliomyelitis chronica, progressive muscular atrophy of the Aran-Duchenne type, amyotrophic lateral sclerosis) syringomyelia can be distinguished by the presence of dissociated disturbances in sensation, trophic disturbances of the bones, joints and skin. Also the one-sided and irregular bulbar affection in contradistinction to the last named disease, the increase of the tendon reflexes in the lower extremities in contradistinction to the former two diseases, present important signs in regard to differential diagnosis.

Multiple insular sclerosis, upon longer observation, will nearly always act clinically in a different way than syringomyelia. Especially the disturbances in sensation are usually not permanent; trophic disturbances do not play an important rôle, more extensive muscular atrophies are quite unusual.

Intramedullary tumors may give rise to considerable diagnostic difficulties. The following factors, among others, are important: the course in tumor is far more rapid than in syringomyelia. The Brown-Séquard symptom complex is a frequent, if only transitory occurrence in tumors. The phenomena of sensory irritation are usually more prominent and pareses or paralyses develop far more quickly than in syringomyelia. Bladder and rectal disturbances, also, are found early. Bulbar disturbances, if they be present, are, as a rule, a very bad sign, whereas in syringomyelia, bulbar symptoms may persist for many years.

The **prognosis** of the disease is quoad vitam not unfavorable, since even when bulbar affections exist, life is usually prolonged for many years. Death is caused, as a rule, by intercurrent diseases.

Treatment is more or less powerless to cope with the disease. It is asserted that use of the X-rays on the spinal column, especially in cervical syringomyelia, may check the progress of the disease. I have not as yet seen any such effect on the progress of the affection.

Prophylactic measures are important. The patients must not expose themselves too much to high or low temperature, since there is danger of burning or freezing. Severe physical work should also be avoided as much as possible, because of a tendency in the affection to the formation of hæmatomyelias and because of the possibility of spontaneous fractures. The use of hot baths may bring about an acute exacerbation of the affection.

The muscular rigidity is influenced favorably by protracted lukewarm baths. The addition of treatment by radium seems to me useful, likewise the use of the baths at Gastein.

If pains be present, the use of the modern antipyretics, especially of pyramidon, citrophen or aspirin, is of service.

Arthropathies in case of suppuration or of too much effusion demand surgical treatment.

5. HÆMATOMYELIA

BY

HERMANN SCHLESINGER (Vienna)

Hemorrhage into the spinal cord is a much rarer affection than hemorrhage in the brain. We differentiate between primary and secondary hemorrhages of the spinal cord, the latter of which takes place into a tissue that has already been changed (for instance, tumors, inflammatory processes, etc.). Hæmatomyelia preferably affects the gray matter; the posterior horns are more frequently injured than the anterior horns. The hemorrhage spreads lengthwise in the spinal cord, may even occupy a large part of the medulla (tubular hemorrhage), may occupy one or both sides of the cord, may injure the anterior horn or the posterior horn alone, while the lateral columns are usually spared (*Minor*). The localization and the spread of the hemorrhage, might, as I assumed years ago, be dependent upon the loose, yielding structure of the gray parts of the spinal cord. Later experimental investigations by *Goldscheider, Flatau* support this view. The hemorrhages may take place in any part of the spinal cord; they are most frequently found in the enlargements. The intrameningeal hemorrhages are mostly of less clinical importance than the intracordal, as *Thorburn* and *Stolper* have shown.

Of the **ætiological factors traumatism** is the most important (for about 9/10 of the cases, according to *Oppenheim*). According to Stolper, forced flexion of the head forwards may cause a hæmatomyelia (for instance, diving head foremost). In newly-born infants hæmatomyelia was seen often after *Schultze's* method of resuscitation of still-born children and difficult deliveries (*F. Schultze, Litzmann*). In children, after forced (*Lorenz*) reduction of congenital dislocation of the hip-joint, hæmatomyelia of the conus was observed (*H. Schlesinger*). After forcible muscular exertion, spinal hemorrhages have been observed, but a predisposition (for instance, hæmophilia) seems to be essential. In typhoid, hæmatomyelia has been observed several times, thus by *Curschmann, A. Schiff*. I saw the disease associated with morbus maculosus Werlhoffii, other authors with pernicious anemia, etc. Spontaneous hæmatomyelias are exceedingly rare, but undoubtedly occur. One of my patients developed it, while working on a farm, without traumatism.

Symptoms.—They vary a great deal according to the spinal level and to the extent of the hemorrhage. Common to all forms is the sudden onset with the phenomena of an interruption in spinal conduction. The maximum of the lesion, i. e., the greatest extent and intensity of the phenomena are

reached, usually, in the first few hours of the disease, far more rarely in the first few days. Part of the symptoms, which are not at once visible (muscular atrophies, trophic phenomena in the skin, etc.), are produced by the lesion, once it is present, but need a certain amount of time for development. The predilection (so striking anatomically) for the gray substance evokes very peculiar symptom groupings, to which Minor especially has called attention. Very often *syringomyelic dissociation of sensibility*, i. e., analgesia and thermo-anæsthesia, with other qualities of sensation retained or but slightly injured, are demonstrable in smaller or larger portions of the surface of the body.

If the lesion is in the sacral cord, bladder and rectal disturbances and occasionally anomalies in the sexual sphere, are added to the disturbances of cutaneous sensibility in the perineum and about the anus.

With injury in the lumbar and lower dorsal segments of the cord, there are usually added also paralyses of the musculature of the legs (severe atrophic paresis in one of my cases) and the tendon reflexes may show important disturbances (loss or increase). Most frequently, the hæmatomyelia is located in the cervical segment and evokes atrophic paralysis of the upper extremities, whereas in the lower extremities, spastic paresis holds sway. The extent and degree of muscular atrophy vary extraordinarily. When the hæmatomyelia is deep-seated (lower cervical enlargement) the cervical sympathetic of the same side is often affected (disturbances in sweat, oculo-pupillary phenomena, vaso-motor disturbances in the face).

The Brown-Séquard symptom complex is developed with remarkable frequency. As *Minor*, *Oppenheim* and others have shown, the disturbance in sensation is partial; indeed, it may be for pain and temperature in one leg, while the other is spastically paralyzed, and the upper extremity on the opposite side is more or less atrophic.

Another form corresponds to the "classic" type of syringomyelia; there is atrophic paralysis of an upper extremity and partial disturbance of sensation in the same extremity.

Of irritation phenomena, initial pains in the back must be mentioned, which, however, are rarely overpoweringly severe.

Course.—The disease attains its height, as I have already remarked, in the first hours (*Kienboeck*). If serious complications exist (injuries to the vertebral column, lesions of internal organs), these, as a rule, cause death. Hæmatomyelia, per se, even when situated high, is usually not fatal. If observation be over a longer period of time, one sees a more or less rapid retrogression of the symptoms, which, occasionally, is surprisingly complete. In one of my cases (described by *Labin*) lesion of the cervical cord, paraplegia of all four extremities was present, and disappeared completely; the disease finally presented the clinical picture of a conus lesion. All phenomena, which may be explained by blood compression, may retrogress, likewise those symptoms that can be traced to an edema in the substance of the

spinal cord. Finally, those symptoms persist which are caused by complete destruction of spinal-cord substance. The permanent symptoms are disturbances in sensibility, especially those of syringomyelic character, since they are called forth by lesion of the gray axis of the spinal cord. To these are to be added muscular atrophies, which owe their origin to destruction of parts of the anterior horns; in the atrophic muscles, the electric excitability is changed (reaction of degeneration). Real trophic disturbances seem to be exceptional only, in hæmatomyelia. The muscular atrophies may be very slight, and are therefore easily overlooked; this is a condition of special importance in judging medico-legal accident cases (*Oppenheim*).

Diagnosis. Differential Diagnosis.—The diagnosis is, as a rule, made easily because of the acute onset, the usually preceding traumatism, and the beginning retrogression of the phenomena within a short time of the onset of the disturbance.

From a differential diagnostic standpoint the diseases most concerned are syringomyelia (cf. above), and *acute myelitis*. In the latter disease, I have observed a most acute onset, with paralysis in several hours, and death within a few days in three cases, and the literature furnishes similar observations. In these cases, however, there is usually fever, and the spontaneous origin also militates against hæmatomyelia.

The greater prominence of irritation symptoms (pains, rigidity of the spinal column, sensitiveness of the muscles to pressure), particularly when the paralytic phenomena are but slightly pronounced, points to *hemorrhage in the meninges* (Hæmatorrhachis).

In *tumors* accompanied by hemorrhage into the tumorous tissue, the anamnesis, as a rule, points to a longer duration of the disease.

In *poliomyelitis*, the almost exclusive anterior horn symptoms and the onset with fever, make the diagnosis possible.

The **prognosis** of the disease depends, as has been said before, on the complications. It is better, when no bed-sores and no cystitis develop.

The **duration** of the disease may be considerable. I possess anatomic preparations from a case that lasted 15 years (destruction of the sacral cord). I know another case in which hæmatomyelia developed 17 years ago, and in which I have been able to demonstrate the condition (Conus lesion) as stationary for more than 10 years.

Treatment.—The most important therapeutic factor is absolute rest—if possible, for several weeks. *Oppenheim* recommends lying on the stomach or side (marked tendency to bed-sores). Internally, gelatin (2 drams daily in watery solution), calcium chloride (20 grains daily in water) or adrenalin (5 drops of the standard solution 4 times a day) are to be advised. Subcutaneous injections of ergotin may be beneficial. Especially in meningeal conditions of irritation, and in full-blooded persons, leeches are indicated in the region of the spinal column.

If atrophic paralyses develop, frequent lukewarm baths may be used; hot baths are contraindicated.

Treatment by baths at Bad Gastein, Wildbad, Nauheim, Oeynhausen are occasionally useful if the hemorrhage has existed some time (at least 5 to 6 weeks). But in treatment by baths, all the more strongly exciting procedures, and especially hot baths are to be avoided.

6. FOCAL DISEASES OF THE SPINAL CORD DUE TO EXTRA- AND INTRA-MEDULLARY AFFECTIONS

BY

R. FINKELNBURG (Bonn)

I. Traumatic Diseases of the Spinal Cord

(*Changes Due to Crushing* (*Contusions*), *Lacerations and Concussion*)

Whenever the spinal column is injured either by *direct* or *indirect* violence, that is, by a *blow*, *contusion*, *by crushing* or by traction upon it or by a fall upon the head or the buttocks, the spinal cord may be injured also. This will be particularly the case if, as a result of fracture of the body of a vertebra, or of vertebral dislocation, there are *permanent changes in the position* of an entire vertebra or of any of the parts thereof. The spinal cord may show *total or partial* contusions and lacerations, *root lesions* with or without hemorrhages into the membranes. But also after only *momentary displacement* of one or more of the vertebral bodies, even if the vertebra has returned to its natural position upon relaxation of the force influencing it, severe *pressure lesions* of the cord may be caused.

The seat of a *dislocation and distortion* is most frequently the cervical portion of the spinal column; the former arises from forced *flexion*, with or without simultaneous rotation of the neck, the latter not infrequently from violent stretching of the spinal column. In isolated vertebral fractures, it is a question either of so-called *compression fractures*, caused by destruction of the body of a vertebra by forces exerted upon its axis, or of fractures in the vertebral arches, which but rarely give rise to an injury of the cord and the roots. Fractures in the body of a vertebra occur mainly in the region of the lower dorsal and lumbar vertebræ. Isolated lesions of the inter-vertebral discs are rare; they frequently accompany dislocations and fractures of the vertebræ.

Apart from the serious injuries of the spinal cord resulting from *permanent* or *transitory compression*, caused by vertebral injuries, even after *simple concussion* to the spinal column, coarser or finer spinal-cord changes may arise. The former consist in extra- or more frequently intra-medullary hemorrhages and in softening of the substance of the spinal cord with possibly additional *chronic myelitis* of the white and gray matter at a later period (cf. Fig. 81).

Slight *concussional injuries* in the form of delicate changes in the ganglion cells and nerve fibres, as may also be evoked independently in experimenting on animals by concussion upon the spinal column, frequently disappear entirely, after leading at first to serious phenomena of functional loss. But occasionally in connection with such a *commotio spinalis*, even if the incipient functional disturbances have disappeared, there develop *insidious degenerations of the spinal cord* in the form of a chronic myelitis. It is of special practical importance, that after commotio spinalis, in which the *brain* also is usually *co-affected*, together with organic changes, but also very frequently without them, pure *functional disturbances* of the nervous system may appear—*traumatic neurasthenias and hysterias*. These commotion neuroses were formerly spoken of as "*railway spine*," since they were observed especially after railway collisions.

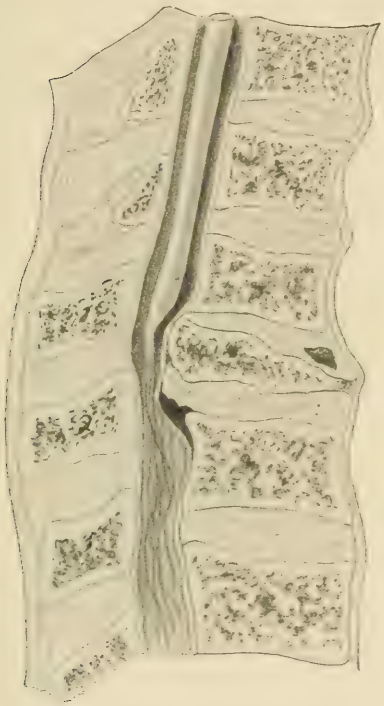


Fig. 81.—Fracture of the first lumbar-vertebra due to compression. (*After Th. Kocher.*)

Anatomic Findings.—After severe crushing there are found, in addition to hemorrhages in the membranes and substance of the spinal cord, marked flattening, diminution, or even complete division of the spinal cord. This is soon followed by a *hemorrhagic softening*, in the further course of which the destroyed and liquefied remnants of the cord are absorbed and partly taken up by granular cells.

In the vicinity of the lesion there appear *inflammatory reaction phenomena;* the glia cells and fibres swell and multiply; the vessels are surrounded by granular cells, and occasionally numerous round cells are to be seen in the area of destruction. The outcome is a *cicatricial focus*, consisting of glia tissue with thickened vessels, occasionally, also, with the formation of larger or smaller cysts. Ascending and descending degenerations are added to the picture.

Phenomena of the Disease

Immediately after every severe injury of the spinal cord, there appears, during the first effects of shock, the *symptomatic picture of a complete interruption in conduction*, even when the spinal cord is but partly injured. It is therefore only after the *first shock phenomena have passed away*, that we can

approach the therapeutically and prognostically important question: is there a *total* or only a *partial* transverse lesion, is there *present crushing* or merely *compression* of the spinal cord?

1. *Complete Transverse Lesion*

The most important sign of a *complete* transverse lesion is found in the *complete paralysis* of the musculature of the body below the point of the lesion, which appears immediately after the injury and remains *permanently*. At the same time motor irritation phenomena, twitchings, spasms in the paralyzed region are wholly lacking, while they may appear near the upper boundary. Even when the seat of the lesion is in the upper segments of the spinal cord, the paralysis of the lower extremities, as a rule, is *flaccid*, and not only during the period immediately following the injury, but usually afterwards as well.

The *tendon* as well as the cutaneous reflexes are completely lost in total lesions above the lumbar segment, not only immediately after the injury, but often *permanently*. But permanent loss is by no means always the case. The theory of *Bastian*, that *every total transverse lesion* of the spinal cord brings with it a *permanent loss of the tendon reflexes* below the point of injury, is disproven by a number of indisputable observations, in which, although complete division of the spinal cord was anatomically demonstrated, the tendon reflexes remained long after the injury and occasionally even showed an increase.

The *loss of sensation* in all its qualities is *complete*, up to the level of that region of the skin which is supplied by the destroyed segment of the spinal cord. The sensory disturbances extend only then farther upwards than would be expected, judging from the position of the injured segment, if by the crushing the root-fibres originating in the higher segments and passing down through the spinal canal are also destroyed.

This, however, is by no means always the case, since in an injury, which strikes roots and cord simultaneously, the former are frequently less injured. *Sensory irritation phenomena* in the form of radiating pains, hyper- and paræsthesias occur near the *upper limit of the disturbance in sensation*, never, on the other hand, within the sensorily paralyzed region.

Disturbances of the bladder and bowels are always present and decided. There is, at first, retention of urine, which later is followed by a periodic reflex evacuation of the bladder. Bed-sores appear sooner or later.

2. *Partial Lesions of the Cord*

In these, the motor phenomena of paralysis appear, as a rule, in the foreground of the disease picture in contradistinction to the usually less pronounced sensory disturbances. In lesions, in which only a part of the transverse section is affected, we see, a short time after the injury, the *motor*

paralysis, which, at first is total, become transformed into one that is incomplete. At the same time, according to *Kocher's* observations, the parts of the body lower down, that is, especially the lower extremities, are more severely affected by the paralysis, than the upper parts, like the arms. Another important sign of a lesion of the cord that is only partial is the appearance of *motor irritation phenomena*, of twitchings and early contractures in the regions showing a motor paralysis. If the point of injury is above the lumbar segment, there appears, as a rule, a spastic condition of the musculature and a pathological *increase of the tendon reflexes*, also the *Babinski phenomenon.*

In partial destruction of the cord *sensation* even in pronounced paraplegia may be but slightly disturbed; furthermore, the disturbance may at times be confined to definite qualities in sensation only and does not extend nearly so far upwards as the region of the motor paralysis. The *appearance of radiating pains and paræsthesias in the paralyzed limbs* is, in a certain measure, *characteristic, in contradistinction to the total transverse lesions.* If there are only disturbances in sensation without motor paralysis, this indicates that only a root injury is present.

Not very infrequently, the symptom complex of a *Brown-Séquard* unilateral lesion develops in injuries due to stabbing, shooting, or crushing: spinal hemiplegia on the side of the injury, with later appearing spastic phenomena, and considerable exaggeration of the tendon reflexes; on the paralyzed side, there is found, in addition, a *hyperæsthesia* and *hyperalgesia* of the skin, and at first, *paralysis of the blood-vessels.* In the opposite side of the body, which does not present motor disturbances, sensibility is impaired, with the exception of the sense of position. Pain and temperature sensations suffer most frequently and most severely. This typical picture of Brown-Séquard paralysis is generally blurred because the injury does not as a rule confine itself definitely to one-half of the spinal cord, or, because on account of an added *traumatic myelitis*, the original semilesion becomes transformed into one diffusely affecting the whole transverse area.

Disturbances of the bladder and bowels are frequent concomitants of an incomplete transverse lesion: but they may be entirely absent in spite of pronounced paralytic phenomena. In partial lesion of the cord, retention of urine with completely preserved bladder sensation may be observed; the sensation in many instances may even be *morbidly increased*, thus inducing violent *vesical and rectal tenesmus*, and at the same time, an impossibility of voluntarily evacuating the bladder and bowels. Priapism is no unusual phenomenon in partial lesion; it may, however, be lacking even in total transverse lesion.

For the general ascertainment, as to which of the *main divisions* of the medulla spinalis is affected by complete or partial transverse lesion, the following table gives sufficient data. The exact *determination of the place* which is affected, in the absence of external injuries, depends above all, on

the detection of motor and sensory phenomena of functional loss of definite segments of the spinal cord. These are best shown in the tables of *Edinger and Starr-Bruns*, and the *sensibility schemes of Edinger, Head, and Seiffer.*

TABLE FOR THE GENERAL DETERMINATION OF THE PLACE OF TRANSVERSE LESIONS (*After Schultze*)

	Motor	Sensory
Transverse injuries of the *cauda equina*, cause:	*Flaccid paralyses* in the sciatic region and the sacral nerves only, or also in the crural or obturator region.	*Anæsthesias* in the same areas; in individual parts, also *paræsthesias, hyperæsthesias* and pains.
Transverse injuries of the *lumbar enlargement*, cause:	According to the place of the injury flaccid paralysis in the sacral nerves only, or also in the lumbar (crural, obturat).	Like the above.
Transverse injuries of the *dorsal segment*, cause:	Paralysis of the lower extremities and individual parts of the trunk, in lesions of the uppermost parts also *oculopupillary symptoms.* Mostly *spastic* paralysis.	*Anæsthesias* of the lower extremities and symmetrical parts of the trunk. At the upper boundary also hyperæsthesias and pains.
Transverse injuries of the *cervical enlargement*, cause:	Spastic paralysis of the lower extremities; paralysis of the muscles of the trunk; partial or complete flaccid or atrophic paralysis of the muscles of the upper extremity.	Anæsthesias of the lower extremities of the trunk up to the height of the shoulder, partly or entirely of the upper extremities. Hyperæsthesias and pains.
Transverse injury of the uppermost *cervical segment*, causes:	Usually immediate death, or if the injury is at the lower part and with paralysis of the phrenicus, fatal termination after days or weeks. Radiating pains especially in the n. occipit. major.	
In respect to *bladder* and *bowels* and sexual function.	In respect to *reflexes.*	*In respect to trophic disturbances.*
Severe paralysis. Later *cystitis.* Impotence.	Loss of the cutaneous and tendon reflexes in the nerve areas concerned.	Reaction of degeneration and wasting of the muscles. *Decubitus:* trophic disturbances of the skin, joints and bones.

Bladder, bowels, and sexual function.	Reflexes.	Trophic disturbances.
The same as above. Impotence.	The same as above.	The same as above.
The same as above. Impotence.	Retention or exaggeration of the tendon and cutaneous reflexes. Sometimes, also, absence of the tendon reflexes.	No reaction of degeneration. Often marked diminution of the electric excitability. Decubitus.
The same as above. Impotence; sometimes priapism.	The same as above.	Reaction of degeneration in the muscles of the upper extremities otherwise as above. Often rise in temperature.

Diagnosis.—After the first effect of the shock is over, differentiation between a total and a partial transverse lesion is, as a rule, possible without much difficulty. In the total cases, *permanent, complete, motor and sensory paralysis* is present without any symptom of irritation in the paralyzed region, while in the partial cases, *incomplete paralysis*, only *partial* sensory disturbance, motor and sensory *irritation phenomena* and increased vesical and rectal tenesmus are found.

On the other hand to determine whether a break in conduction depends upon a complete or a partial *destruction of the nerve substance*, or whether it is merely a matter of the *effects of a simple compression*, is more difficult and under some circumstances impossible. It is certain that a total interruption of conduction may even be produced by a lesion causing pressure. Likewise, concussion of the spinal cord, through hemorrhages into its substance, may cause, even without *greater* anatomic changes in it, severe functional disturbances, which may simulate a contusion. In either case rapid improvement frequently occurs. As a rule, definite differentiation between a crushing of the cord and simple compression is made possible only by long observation. The longer duration of the symptoms of functional loss without signs of improvement makes probable the presence of *irreparable lesions of the cord.*

Prognosis.—This is *serious* in all injuries of the spinal column, in which the spinal cord is affected. *Total* transverse injuries are *fatal* without exception, if not in direct connection with the traumatism, then later as a result of the unavoidable complications of decubitus and cystitis. Nor is the prognosis in partial lesions of the cord quoad vitam always favorable, since a long time may elapse before there is functional restoration and especially before adjustment of the disturbances with respect to the bladder and bowels, while the patient may meanwhile die in consequence of a decu-

bitus or of a cystitis. The prognosis is made still worse by the fact, that hand in hand with the improvement of the at first severe phenomena of paralysis, obstinate irritation symptoms are caused by the sclerotic processes following closely upon the destruction of tissue, which may disturb considerably the ability to use the limbs, which otherwise has been fortunately regained.

Treatment.—This in the days immediately following an injury will have to be restricted to preventing further displacement of the broken or dislocated vertebræ, by careful placing in a recumbent position and the avoidance of all movements. Beside this all measures to prevent a decubitus are of the greatest importance. Regarding *operative* results and especially as to the *proper time* for operative interference, views differ considerably. The majority reject an early operation (before the sixth week), since in the incipient period the phenomena caused by the commotion and intra-medullary hemorrhages can not be distinguished from those due to crushing, contusion or compression.

If the symptoms of a total transverse lesion persist, operative interference is useless. In cases of incomplete interruption the spontaneous improvements are often very extensive. But if no progress is clearly discernible after some weeks, this indicates rather that the spinal cord is injured not only by a *transitory lesion due to pressure* but that a *permanent compression* from detached bone splinters, or displaced vertebræ, is present.

Since in such cases there is a possibility of effecting an improvement or a cure by removing the pressure, an operation is advisable.

II. The Pressure Paralyses of the Spinal Cord

As an important cause of either quickly or slowly appearing *compression paralyses* of the medulla spinalis, are to be considered not only the *tumors of the spinal cord and its membranes*, and the chronic thickenings of the spinal coverings, which similarly to *pachymeningitis cervicalis hypertrophica*, act locally like a tumor, but also and more especially the diseases of the individual *vertebræ*.

We observe *suddenly* developing pressure paralyses after *vertebral fractures and dislocations*. Among the changes in the spinal column that effect a *slow* gradual compression of the spinal cord, *vertebral caries* plays the main rôle. In the great majority of cases it is *tuberculous;* but other forms are known—spondylitis following *traumatism*, *typhoid*, *pneumonia*, and an *acute osteomyelitis* of the vertebræ.

More rarely than by tuberculous caries, the spinal cord is injured by *neoplasms of the vertebræ* of a primary or a metastatic nature or by malignant tumors, which starting from the neighborhood of the spinal column, extend to it and the spinal cord. *Syphilis* also can cause compression of the cord by *exostoses* upon the body of the vertebra and its processes.

(a) Caries of the Spinal Column, Spondylitis Tuberculosa

Tubercular vertebral disease occurs at all periods of life, but especially in childhood and adolescence. It may appear as the only clinical symptom of tuberculosis; but usually, other signs of an *arrested tuberculosis*, or of one *still active* are demonstrable. Traumatisms of the spinal column may play a rôle in the origin of caries, in that *localization* of tubercular processes in the vertebræ is favored by a fall, blow or contusion in tuberculous individuals.

Pathological Findings.—The seat of the tubercular granular tissue, which slowly consumes the substance of the bone, is particularly in the body of the vertebra; less frequently the process begins in the vertebral joints, intervertebral discs or arches. If the vertebral bone, destroyed by caries, is gradually compressed by the weight of the body, the consequence is a separation of the spinous processes, a *sharp-angled projection or lateral divergence* of the spinous process of the diseased vertebra, a so-called *Pott's kyphosis*. This may appear quite suddenly, if through a direct traumatism, or through severe physical exertion, as in lifting loads, etc., the diseased vertebra suddenly breaks.

The tuberculous process does not, as a rule, remain limited to the bone; after a time there is an accumulation of cheesy purulent substance between it and the dura and the latter itself undergoes inflammatory changes, a *peripachymeningitis* develops; from the pressure of the dura mater, as a result of the cheesy substance and its *inflammatory thickening*, the spinal cord and its roots are, as a rule, considerably compressed. A direct transference of the tubercular inflammation to the spinal cord itself through the dura or by the way of the root bundles and blood-vessels, rarely takes place.

As a rule, a distinct *constriction* and *diminution* in size of the spinal cord is to be seen at the point of compression. A microscopic examination may disclose comparatively slight changes in tissue, even though during life serious disturbances in conductivity existed. In such cases groups of *swollen axis cylinders* and *medullated sheaths* with incipient *phenomena of degeneration* may be seen in the midst of well-preserved nerve fibres; the *glia tissue* also shows *infiltration phenomena* and seems thickened. The absence of inflammatory changes proper is emphasized by the majority of authors. If the process has gone farther, wide-meshed spaces have been formed through the degeneration of nerve fibres, which are filled with granular products of degeneration and granular cells. In the later stages after the pressure has been exerted quite a long time, it comes through glia proliferation to the formation of sclerotic focal lesions in place of the destroyed nervous tissue. Secondary *ascending* and *descending degenerations* may follow in their wake.

Phenomena of the Disease

To the early symptoms of a developing caries, there belongs not infrequently a dull *back-ache* caused by the vertebral disease; it is usually localized

to a definite place and is often felt only after *motion*, through *concussions*, and by *direct pressure*. A symptom that results from the pains that appear upon motion, is a certain *stiffness* of the spinal column, which is especially apparent upon bending, as the patients will sink upon their knees to spare the spinal column. But this local painfulness of certain vertebræ increasing upon pressure, is by no means a regular phenomenon; it is occasionally absent even when a *sharp-angled kyphosis*, or a lateral divergence of a spinous process can already be demonstrated; they are the *most important symptoms* of a caries. This change in form of the spinal column is most prominent in the dorsal portion, whereas in a caries of the cervical and lumbar portions, it may be recognized at times only as a flattening of the normal curvature. Since the vertebral deformity as well as the pains may be *absent* during the *entire course* of the disease, the possible proof of the *burrowing of pus* from the diseased vertebræ is of great diagnostic importance. These so-called *burrowing abscesses* in caries of the upper cervical vertebræ appear occasionally in the space between the posterior pharyngeal wall and the spinal column—in caries of the lower cervical and dorsal vertebræ, following the course of the psoas muscle in the inguinal region; a rupture into the thoracic cavity has also occasionally been observed.

Root and Spinal Cord Symptoms

These are not infrequently wholly absent, even in the presence of clinically pronounced symptoms of caries—pains, stiffness, gibbosity—which is not at all remarkable, since it is not the bending of the spinal column that causes the appearance of compression phenomena, but only the *narrowing of the spinal canal*, through the thickening of the dura and the accumulation of cheesy tuberculous masses between the dura and the bones, which need not be present in every case. But it happens also that even though the vertebral phenomena are present only in slight degree, already pronounced spinal disturbances are demonstrable.

As the first sign of compression *root-irritation symptoms* appear, which are of a *neuralgic* nature and according to the seat of the injury, may appear as uni- or bilateral *occipital*, *branchial*, *intercostal*, *crural or sciatic neuralgia*. Not infrequently there exist, at the same time, paræsthesias of great variety. Sensory phenomena of functional loss, *hypæsthesias and anæsthesias* appear only later, when the nerve roots are destroyed to a greater extent. Atrophic paralyses of definite muscular regions, through the destruction of the appertaining anterior roots, also develop, only later as a rule.

If in the progress of the disease, the cord itself is injured by the increasing pressure, this is shown as a rule by the *increased exaggeration* of the *tendon reflexes* lying below the place of the compression. Even before the appearance of paralytic phenomena, *patellar and ankle clonus*, and the *Babinski sign*

are present. The *motor paralysis* of the legs, which next appears either quickly or slowly in a caries situated above the lumbar segment, is at first flaccid and later spastic.

Disturbances in sensation may be *absent* for a strikingly *long time* in caries, or be but slight, even though a pronounced motor paraplegia is already present. An attempt has been made to explain this by the fact that the indurated *peripachymeningitis* compresses especially the pyramidal tracts from the front and the sides, while the *posterior columns* are *spared* more, and that just as in lesions of the peripheral nerves, resulting from pressure, the sensory fibres have greater ability to resist the pressure than the motor fibres. But in a more severe injury of the cord from pressure, a decided sensory disturbance, affecting all qualities, will gradually appear in caries also—a disturbance that occasionally is bounded by a rather sharp upper limit, and extends to the region of the roots, which originate from the diseased segments of the spinal cord. In rare cases, the *Brown-Séquard* symptom complex has also been observed in caries (*Oppenheim*).

Disturbances of the bladder and bowels are occasionally seen *early* along with exaggeration of the tendon reflexes. They are never absent, when the motor and sensory phenomena of paralysis indicate a more extensive transverse lesion. In such cases, bed-sores also may easily develop.

The *general symptomatic picture* of caries (which has just been discussed) will exhibit certain characteristic peculiarities, according to the seat of the carious vertebral change in the upper and lower cervical portion of the vertebral column and in the lower dorsal and upper lumbar vertebræ. These peculiarities we shall have to discuss briefly.

1. In caries of the *upper cervical vertebræ* and of the *atlanto-occipital joint*, beside *pains and stiffness in the back of the neck*, a *neuralgia of the occipital nerves* on one or both sides is the most important early symptom. In all changes of position of the upper part of the body, the patient supports his head with his hands (*Rust's symptom*). *Oppenheim* mentions also the occurrence of paralysis of the *accessory nerve* and *unilateral atrophy of the tongue;* upon compression of the medulla oblongata *bulbar symptoms* are added; where pressure lesions are produced in the upper cervical segments, *phrenic phenomena* will be noted in some cases. There are, besides, *spastic paresis of the arms and legs* and paralyses of the musculature of the trunk, as well as sensory disturbances extending to the neck.

2. If the cervical enlargement is affected in disease of the lower cervical vertebræ, this may be recognized by the fact that beside spastic paresis of the legs and weakness of the musculature of the trunk, a *flaccid* paralysis of the muscles of the arm and hand develops with *muscular atrophy* and electrical reaction of degeneration.

3. A caries of the eleventh and twelfth dorsal vertebræ and of the first lumbar vertebra results in a pressure lesion of the lumbar enlargement.

The *paralysis of the legs* is, therefore, from the first, *flaccid and atrophic* with signs of electric reaction of degeneration; at the same time, the *tendon reflexes are markedly diminished or even lost.* The sensory disturbance remains restricted to the legs; bladder and bowels usually show considerable disturbances.

Diagnosis.—The diagnosis of a compression paralysis is easily made, if besides the *root and cord symptoms*, which indicate a progressive transverse lesion, a *deformity of the spinal column* with local sensitiveness to pressure, may be demonstrated. Then only the further differential diagnostic determination is necessary as to whether a tubercular process is the basis of the vertebral affection, or if a tumor of the spinal column, especially a carcinoma is to be assumed. The exclusion of the usually rare vertebral disease following traumatism, typhoid, acute osteomyelitis, will, as a rule, be easy by taking into consideration the ætiological factors.

The matter may be far more difficult if, as not rarely occurs, symptoms that point to a disease of the spinal column are absent for a longer time and only the signs of an increasing transverse disease of the cord are present. Under such circumstances positive differentiation between *tumors* of the spinal cord, the spinal membranes and *myelitis* having a *syphilitic* or a *tubercular* basis on the one hand, and a caries or a *vertebral tumor* on the other, is absolutely impossible at first and becomes possible only after long observation.

For the diagnosis in favor of caries, in the first place, the proof of other signs of *recent or arrested tuberculosis* (*fever*, *enlarged glands*, *affection of bones*), must be considered. Occasionally also it is possible to establish destruction or displacement of the vertebræ by means of the X-rays. Furthermore the ascertainment of burrowing abscesses may be decisive; value has also been laid upon the greater stiffness of the spinal column in caries in contradistinction to tumors of the spinal cord. Even if it is certain, that larger tumors of the spinal membranes may run their course without special hindrance to the motility of the spinal column, this holds good actually only for tumors of the dorsal part, while those in the region of the cervical segment occasionally cause a considerable stiffness in the neck.

The **differential diagnosis** in regard to a *meningomyelitis syphilitica* must be especially based on the fact that in this disease, the symptoms show quite a *striking variety* and that, as a rule, the picture of this disease points to *multiple*, *spinal* and also *cerebral foci.* In a doubtful case the success of a course of specific treatment may shed light on the nature of the disease.

For the distinction between a *spondylitis tuberculosa* and *vertebral tumors*, one must consider that the latter usually attack and infiltrate several adjacent vertebræ and therefore not infrequently cause a simple *sinking into each other* of the vertebræ (changes easily seen on the Roentgen plates), and a *diminution in height* of the patient; also, as a rule, the developing gibbosity is

less angular than in caries, since several vertebræ participate in the deformity. The age may afford a further important point, inasmuch as in the young, the assumption of a vertebral carcinoma has in itself little probability, while, on the other hand, in more advanced age with a more complete loss of strength, a suspicion of vertebral cancer seems justified and makes it advisable to seek for a primary cancerous focus.

Prognosis and Course.—The tuberculous bone process, which may extend over years without spinal phenomena necessarily occurring, may at any time come to a standstill and recovery. The same holds true of the spinal paralytic symptoms, which even after persistence for *months and years* may *completely disappear* again. The course of the disease, however, is but rarely so favorable. In the majority of cases of caries, with compression of the spinal cord, death results, whether through progress of the tuberculosis in other organs or by the unavoidable results of a decubitus or cystitis in advanced paralysis. Sometimes the phenomena of the spinal cord disappear only in part, as after healing of the bone disease and the inflammatory processes in the membranes the *sclerotic* changes in the spinal cord have already advanced so far, that a compensation of the functional disturbances is no longer possible. In such cases as permanent signs of the former compression, there are more or less pronounced *spastic pareses* in the extremities, possibly combined with *disturbances in sensibility and of bladder function.* A difficulty in micturition was, in one of my cases, after the disappearance of all the other spinal symptoms, for years, the only remaining really disturbing phenomenon. The prognosis is the more favorable, the less the phenomena of compression have advanced at the beginning of the treatment and the younger and stronger the patient. It is remarkable that relapses, especially after traumatism, may appear at any time and that even after persistence of a simple gibbosity for decades, spinal symptoms may still appear occasionally afterwards.

Treatment.—The most important thing in every caries with marked phenomena of compression is a position assuring *absolute rest* for the diseased spinal column. Even by simply *lying on his back* in bed for a long period of time, the patient may occasionally bring about a complete cure from severe compression paralysis. In addition the most careful general nutrition and care of the skin is essential for the prevention of bed-sores (smooth mattress, washing the places where pressure occurs several times daily (alcohol and water), air or water cushions).

Very rapid *improvement* of the paralytic symptoms is observed after *careful extension* of the spinal column. This is most easily performed in caries cervicalis, as the pull upon the head is effectually exerted by *Glisson's* suspensory apparatus with light loads (3 to 4 lbs. at the beginning, and never exceeding 12 to 15 lbs.), the opposite pull being effected by the weight of the body, the bed being elevated at the head. Occasionally, after using exten-

sion, a deterioration is observed, so that a return to simple rest upon the back without further stretching becomes necessary.

If the pressure phenomena are but slight from the first, or if the paralysis has disappeared by rest in bed, and the general health and strength of the patient is excellent, ambulatory treatment with a well-fitted *plaster-of-Paris corset* and the usual supporting apparatus by which the diseased vertebra is relieved from the weight of the parts of the body lying above it may be resorted to (*Sayres* Jury mast, the head supports of *Nebel*, *Heusner*, *Schede*). Ambulatory treatment has the great advantage that through it the patient may be enabled to live in the open, in the fresh air (seaside resort) and thus through the movement and exercise of his musculature, a better circulation, an increase in appetite, and a better general condition are attained.

Operative interference has been suggested and attempted in such cases, in which the compression paralysis remains the same, or even increases under the usual extension treatment. It is followed in almost every instance by an immediate favorable result since the phenomena of paralysis improve immediately after the tuberculous focus is removed. As a rule, however, the improvement does not persist, and the resection of the vertebral arches—in the case the tuberculosis is not restricted to these alone—has many more failures than successes to its account. At any rate, *Schede* considers the operative opening of the spinal canal, as a last resource, not only permissible, but under circumstances even necessary.

The course prescribed by *Calot* for recent and old cases, forcibly to press back under anæsthesia the projecting vertebra and to fix the position so attained by bandages, has been abandoned as being too dangerous.

Of local measures, rubbing with soft soap, and painting with iodine have been frequently used. *Oppenheim* saw a favorable influence, several times, from iodide-iron preparations.

For combating spastic conditions, but also in paralysis without severe muscular spasms, if the patients can be moved, the use of salt and brine baths and an eventual "bath cure" in *Nauheim* and *Kreuznach* are to be recommended.

Electric treatment, at best, should be confined entirely to old cases that have run their course, with flaccid paralysis and atrophic muscles. Direct galvanic treatment applied to the back is productive at times, according to *Oppenheim*, even in old cases, of favorable effects.

(b) Carcinoma and Other Tumors of the Vertebral Column

The not infrequent *vertebral cancer* is nearly always metastatic, and is found relatively frequently in *carcinoma of the breast*, but the abdominal organs must also be taken into consideration as the place of origin. The connection between the vertebral spinal cord phenomena and cancer remains quite often unsuspected for some time because the *primary cancerous focus*

at first causes few pronounced clinical symptoms, or, as in carcinoma of the breast, under certain conditions, it has been removed by operation some time before the appearance of the disease of the spinal cord. As a consequence, doubts as to its nature may exist for some time. Vertebral cancer, like most of the other tumors, which, beginning either in the vertebræ themselves or in their neighborhood, attack the vertebræ, effects, as a rule, *several vertebral bodies*. Owing to the fact that the tumors extend to the vertebral processes, and also infiltrate the soft parts enveloping the spinal column, there arise occasionally large, easily palpable excrescences on the spinal column. As a rule, however, this is not the case, and as a direct consequence of vertebral destruction, we find only a *sinking of one body into the other* (Fig. 82). Besides carcinoma, which is by far the most frequent and therefore the most important tumor of the spinal column, there have been described *sarcomata*, *osteosarcomata*, the rare *enchondromata*, the multiple appearing *myelomata*, also *gummata and echinococcus cysts*.

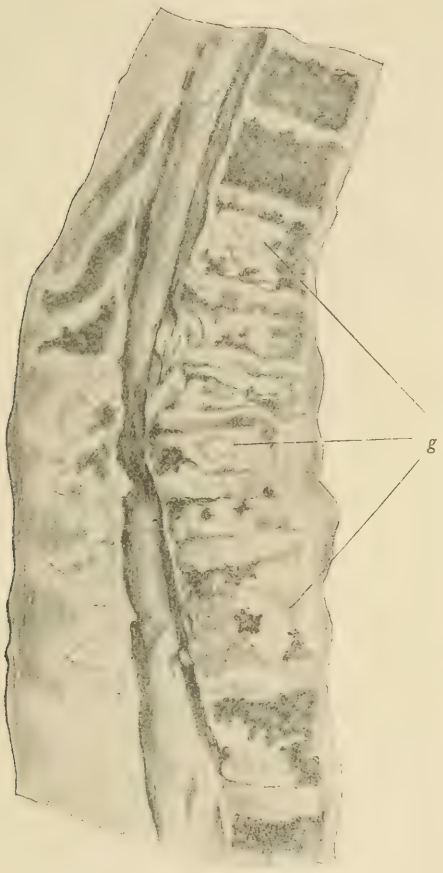


FIG. 82.—Carcinoma of the vertebral column with pronounced and rather uniform compression of several vertebræ; (*g*) destroyed by the tumor and a narrowing of the vertebral canal due to this. (*After Schlesinger.*)

Syphilis also leads to the formation of exostosis on the body of the vertebra and its processes.

Symptomatology.—The disease picture shows great similarity to that of a *spondylitis tuberculosa*. In the typical cases, there are found besides obvious changes in the spinal column with local vertebral pain, which is increased upon motion, *root irritation symptoms and phenomena of compression of the cord*. Demonstrable changes in the bones, however, may be absent in carcinoma at a time when root and cord symptoms are already pronounced. As an important bone symptom, there appears occasionally a gradual *lessening of the length of the body* through the sinking together of several vertebræ or a *gibbosity*, which because of the disease in several vertebræ, is usually less pointed than in caries. More rarely it comes to the development of large palpable thickenngs of the vertebræ.

The pains, not only the local pains, but the neuralgic pains produced by root compression as well, are in vertebral tumors, as a rule, *extraordinarily violent* in contrast to those due to caries. The spontaneous pain is not rarely very severe, though the local tenderness to pressure, which even in extended vertebral carcinoma may be restricted to several vertebral spinous processes, is but slight. The cord symptoms, *motor* and *sensory* paralyses, and *disturbances in bladder and rectum*, develop rather quickly upon occasion, when a carcinoma after breaking through the dura, invades the cord. With advanced changes in the bones, a slight traumatism may suffice to give rise to a sudden collapse of the vertebræ and an *acute paraplegia*.

Since in the paraplegic limbs, the pains usually persist with great violence *Cruveilhier* long ago spoke of a *paraplegia dolorosa* specially in vertebral carcinoma.

The course of the disease is rapid in the majority of cases; especially in sarcomata and carcinomata the disease rarely lasts longer than nine months to one and one-half years.

Diagnosis.—Even if a deformity of the spinal column points to a compression paralysis, differentiation from a caries will not always be easy. The diagnosis of vertebral cancer will have to be based on the *age of the patient*, which justifies a suspicion of carcinoma, upon *general loss of strength*, above all, on the demonstration of a *primary cancerous focus* in other organs and on a history of cancer, extirpation of the breast, etc. On the other hand, youth and the proof of recent or healed tuberculosis speak for a diagnosis of caries. It is important for differential diagnosis, that in carcinomatoses also spinal phenomena may be observed, which depend upon a disseminated myelitis. In the absence of definite changes in the bones, and of stronger root irritation symptoms, with the demonstration of a primary tumerous focus, we must reckon with this possibility. An X-ray investigation, which should always be tried as soon as possible, will be of further assistance in these cases also.

Treatment.—If there be a suspicion of a *syphilitic* affection of the bones, with the formation of an *exostosis* leading to compression, specific treatment is to be begun, which in a series of cases (*Oppenheim, Leyden, and others*) led to *complete recovery*. Operative interference is only to be advised when the tumor is not a metastatic growth. In the primary tumors of the spinal column, a favorable result can be attained by means of an operation, at least temporarily, as it ameliorates the torturing pains. Recurrence occurs quickly as a rule in consequence of the usually great extension of the tumors over several vertebræ. But according to isolated observations in *osteoma* and *sarcoma* (*Oppenheim*) a cure, which may last for years, seems to lie within the bounds of possibility, so that one must not deprive the patient, if conditions are favorable, of his chance by an operation, though the prognosis is usually hopeless. In the majority of cases, we shall have to limit

ourselves to combating the pains (*morphine, aspirine, pyramidon*), and by a careful recumbent position and avoidance of all unnecessary movements try to attain a permanent relief for the diseased bones.

(c) Neoplasms of the Spinal Cord and its Membranes

Pathological Anatomy.—Tumors of *extra-medullary* development are more common than neoplasms arising in the spinal cord itself. Among the extra-medullary tumors those situated *outside of the dura mater*, if we except those of the vertebræ, are much rarer than the practically much more important *intra-dural* tumors. In the rare *extra-dural* neoplasms, it is usually a matter of *lipomata and echinococci;* among the tumors developing within the dura, *fibromata, sarcomata* and their mixed forms play the most practically important part, since they usually occur in circumscribed form about the size of peas to hazel-nuts and are easily removed by operation; besides these are found intra-dural *myxomata, psammomata,* less frequently *lipomata, adenosarcomata, lymphangiomata, and teratomata. Sarcoma* occasionally appears in multiple form, or it spreads *diffusely* over the membranes of the spinal cord, the individual growths varying much in size.

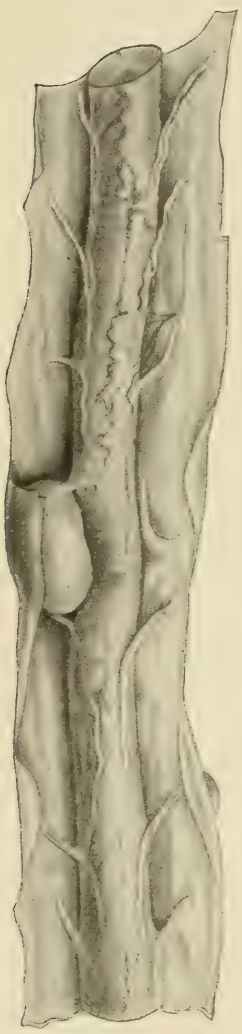


FIG. 83.—Intra-dural tumor, situated opposite the dorsal cord between the 6th and 7th pair of dorsal roots, compressing the spinal cord posteriorly from the right side. Drawing is natural size after cutting open and spreading the dura. (*Author's own observation*).

The greater number of the growths in the spinal cord itself are *gliomata;* they are preferably situated in the cervical and upper dorsal cord and in the upper part of the lumbar enlargement; other neoplasms occurring are *sarcomata, tubercles and gummata.* Multiple *neurofibromata* emanating from the roots of the spinal cord are frequently only a partial symptom of an extensive general *neurofibromatosis* of the nervous system.

Causes.—Excluding the *parasitic* and *infectious* growths we know nothing of the ætiology of the neoplasms and must be satisfied with the assumption of an abnormal congenital predisposition of the tissues. It seems that *traumatisms* are to be considered as exciting factors in the development of the growths or that they are at least factors favoring their quicker progress.

Tumors in the Membranes of the Spinal Cord

Symptomatology.—Since the majority of tumors emanating from the membranes are not malignant

and grow but very slowly, the appearance of symptoms of compression in the substance of the spinal cord is preceded by an earlier *stage of neuralgic* pains, which may extend over months and even years and which are caused by the *irritation* due to pressure upon the *posterior roots*. These radiating pains are localized to one or both sides of a definite nerve region and present the picture of an *intercostal or of a sciatic neuralgia*, or a *neuralgia of the brachial plexus*. Patients frequently complain of an increase in the pain whenever the spinal column is jarred, as in coughing, sneezing, or by pressing upon it. The prodromal stage of pains need not always be present in tumors of the spinal cord membranes; occasionally the pain is present to only a slight degree, or lasts only a short time. *Irritation symptoms* in the anterior *motor roots*, tremors, muscular contractions, spasms occur but *rarely* compared to the sensory irritation phenomena, even where a gradually growing tumor, as shown by its location, must have pressed almost equally on posterior and anterior roots. As a rule, in a compression of the anterior roots, the irritation phenomena are quickly followed by atrophic paralysis in the corresponding muscular area.

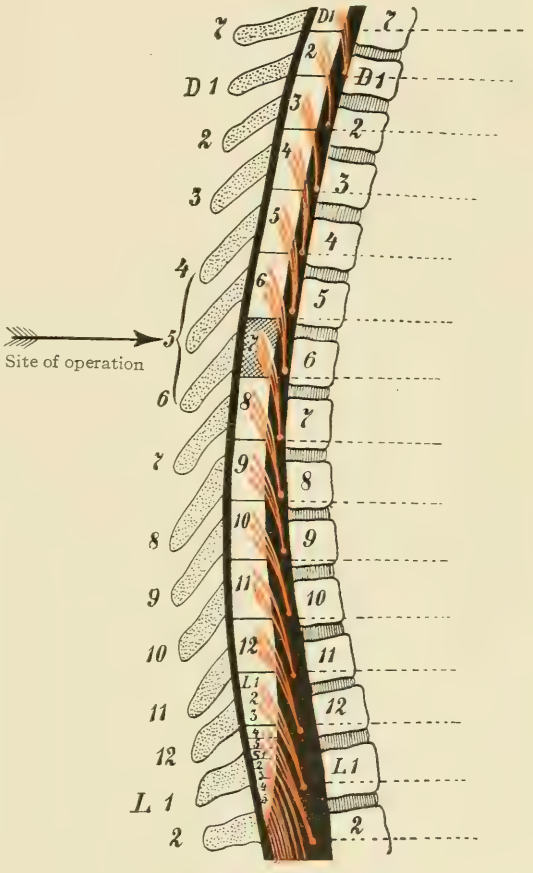


FIG. 84.—Gower's scheme (from the 7th cervical to the 2nd lumbar segment) shows the relation of the spinous processes and of the places of exit of the roots to the vertebral bodies and the relation of the spinal cord segments to the vertebral bodies. The 7th dorsal segment is regarded as diseased.

In the usual location of a tumor in the dorsal portion of the cord, atrophic muscular paralyses caused by pressure lesion of the motor roots fall entirely into the background of the clinical picture, since weakness and wasting of individual intercostal muscles escape detection. Only in cases in which the lower dorsal roots (8–12 pair) are injured by the tumor, can *uni-* or *bilateral paralyses of the abdominal muscles* appear and with the sensory

phenomena of irritation, form an *early local symptom* of considerable diagnostic importance.

A less frequent symptom of irritation, which, however, must be sought for, is a *local tender point* of the spinal column, usually at the height of those vertebræ, opposite to which the tumor is located. This vertebral tenderness is best sought for and tested in this manner: exert strong pressure with the back of the flexed index or middle finger to one side of the spinous processes of the vertebræ. This will be found painful especially on the side which was the seat of the neuralgia at the onset.

As the tumor continues to grow, the signs of a *compression of the cord* are added to the symptoms of root irritation, which persist in most of the cases; a *motor and sensory paralysis* which, as a rule, advances slowly and regularly in the part of the body lying below the place of pressure. The motor paralysis is always *spastic* when the tumor is located, as is usually the case, above the lumbar segment; in the beginning it usually appears as slight paresis in one leg, and later extends to the other side, and even when paraparesis is already present, the side of the body corresponding to the seat of the tumor usually shows the more severe paralytic phenomena.

The *tendon reflexes*, the *increase* of which occasionally is the *first symptom of compression of the cord*, are regularly and considerably exaggerated; *patellar and ankle clonus* are present and frequently the *Babinski sign*, especially in the more markedly paralyzed limb. The *reflexes of the abdominal muscles* are usually lost or weakened.

The *disturbance in sensation* may be limited at the beginning to one leg, because the cord at first is compressed on one side, so that with a simultaneous paresis of the leg corresponding to the side of the tumor, there comes to exist for some time the symptom complex of the *Brown-Séquard unilateral paralysis*. Thus, for instance, *Oppenheim* saw a *thermanæsthesia* in the opposite leg precede the development of the homolateral paralysis in two of his cases. As the transverse lesion increases in size, due to the growing tumor, the *unilateral* sensory paralysis is soon transformed into a *bilateral disturbance;* it may not come under observation until this stage is reached.

In the *typical* cases, the *upper boundary* of the sensory disturbances, which is usually a horizontal line on the trunk, may be established quite accurately upon repeated tests. At this upper margin, sensibility is not completely lost but only *diminished.* This upper *hypæsthetic* cutaneous area gradually passes, as we go downward, over into the region of the skin that is more or less decidedly *anæsthetic.* In some cases, it has even been possible to demonstrate an abnormal sensitiveness in the skin area immediately above the hypæsthetic zone—a *hyperæsthesia.* Especially characteristic and important for the diagnosis of tumor is the determination that the upper boundary of disturbed sensibility remains *in the same place* even

after a longer period of observation, or, at any rate, undergoes only an unimportant move upwards. This depends on the fact, that with a slowly growing tumor, even a slight *increase* in its *thickness* must cause a marked compression in the narrow spinal canal, whereas a *slight increase in length* can cause no perceptible extension of the sensory disturbances upwards by lesions of the roots and the cord.

Disturbances of bladder and bowels are always present as soon as the phenomena of cord compression have developed to a somewhat considerable degree; but even in the beginning of the disease bladder disturbances in the form of increased urinary pressure or of slight difficulty in micturition may make themselves felt.

If the tumor is not located, as we have assumed up to this point in the discussion, in its favorite place in the *dorsal portion* of the spinal cord, but in the region of the *cervical or lumbar enlargement*, or still farther down in the *conus terminalis* or the *cauda equina*, the symptomatic picture, according to the location in one of these regions will present certain peculiarities, important for localization. In compression of the cervical enlargement, we shall see, in addition to *spastic paresis* of the lower extremities after inceptive neuralgic troubles in the course of the nerves of the arm, *flaccid degenerative* paralyses of the arm-muscles develop caused partly by the destruction of the anterior roots, but partly also by injury, through pressure, of the anterior horn ganglion cells of the cervical enlargement. Through irritation or paralysis of the sympathetic fibres, lying in the lower cervical and upper dorsal portion of the cord, there may appear also so-called *oculopupillary symptoms, dilatation or contraction of the pupil* and *palpebral fissure* on one or both sides. Tumors of the lumbar region, because of the fact that the nerve roots are placed close to one another, generally cause very violent and extensive neuralgic pains; also the paralysis that appears later in the legs is from the first flaccid and atrophic and is associated with loss of the tendon reflexes.

The symptomatic pictures evoked by compression of the *conus* and the *cauda* will be discussed in a special chapter.

Intra-medullary Tumors

The intra-medullary tumors in their symptomatology and course frequently differ essentially from the *typical* picture of the tumors of the spinal cord membranes discussed above. In the first place, a pure *neuralgic stage* lasting for any length of time is, *as a rule, absent* in tumors of the spinal substance. Even if with the development of a new growth in the periphery of the cord, in the neighborhood of the membranes and the posterior root zone, pains form the first symptom, nevertheless, *soon afterwards*, or even *synchronously*, appear the signs of a *transverse lesion*, during which a transitory *Brown-Séquard* type of paralysis may be observed. Likewise

the pains excited occasionally during the course of the disease, by tumors of the cord, whether through irritation of tracts within the spinal cord or an accompanying chronic meningitis, are usually only of slight degree, and fall quite into the background of the disease picture.

If a tumor develops from the first in the central portion of the cord, disease pictures may develop, which do *not at all resemble those of tumor of the spinal cord membranes*. In more diffuse extension over a transverse section of the spinal cord, there appears for a long period of time, the picture of a *subacute* or *chronic myelitis transversa*, without any demonstrable cause; in gliomata with simultaneous cavity formation, the phenomena of a *syringomyelia* present themselves; it is precisely in cord tumors that *dissociated sensory paralysis* is found most frequently; destruction of the anterior horns, through tubercles or a sarcoma with rapid extension in the longitudinal axis produces the picture of a rapidly progressing *spinal muscular atrophy*.

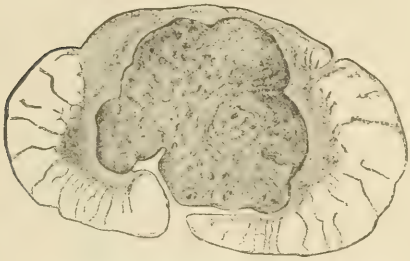


Fig. 85.—Intra-medullary spindle-cell sarcoma. (*After Gowers.*)

For the course of tumors of the cord, it is, moreover, characteristic to a certain degree, that a comparatively *rapid growth* in the *longitudinal direction* upwards and downwards, takes place, and thereby a swifter moving of the boundary of the sensory paralysis upwards is observed than in the tumors of the membranes.

General Diagnosis.—Differentiation of a *tumor of the spinal membranes* from other diseases of the spinal cord not dependent upon compression, which run their course under the picture of a diffuse transverse disease, is not as a rule difficult. Confusion with a *pure myelitis transversa* is prevented by the usually acute onset of this disease without preceding neuralgic disturbance, as well as by the simultaneous demonstration of causes and assisting factors frequently to be considered in acute myelitis, as preceding infectious diseases, anginas, colds, over-exertions, possibly also the existence of fever. For the differential diagnosis from *multiple sclerosis*, which occasionally, for some time presents the symptomatic picture of a chronic transverse myelitis, the chief weight must be laid upon: 1. The *permanent absence* of pains, not only in the beginning of the disease but throughout its course; 2. upon the absence of a sensory disturbance noted evenly and constantly at a certain level, and 3. upon the addition later of other symptoms characteristic of sclerosis, like *nystagmus, intention tremor, changes in the background of the eye*. At any rate, as some experiences have shown, a relatively long period of observation may occasionally be necessary, to exclude a sclerosis with certainty.

Differentiation from a *meningo-myelitis luetica* may not be easy at the

beginning, if relatively violent pains are caused by root irritation. But lues spinalis is characterized by the fact, that no regular, absolutely steady advance of all symptoms can be observed as in the case of tumor, but that in this disease there is frequently considerable *alteration in the violence and extent* of the motor and sensory phenomena of paralysis; furthermore, the syphilis is not as a rule confined to the spinal cord, so that the diagnosis is made easier by *simultaneous cerebral symptoms*. In a doubtful case, the employment of specific treatment will be necessary as a further means of assistance.

We face a far more difficult problem, if it be a question of the practically so important differentiation, whether we are dealing with a tumor of the spinal membranes or with a *compression paralysis* from another cause; through *intra-medullary or vertebral tumors*, *spondylitis tuberculosa*, *pachymeningitis cervicalis hypertrophica* or a circumscribed *meningitis serosa spinalis* (*Oppenheim* and *Krause*).

1. The determination of a *caries* and a *vertebral tumor* is very frequently made very difficult, or even quite impossible, by the fact that in the beginning of the disease, even with the most minute examination of the spinal column, no deformity or localized tenderness can be demonstrated and that examination with the X-rays has had a negative result. A greater degree of *stiffness* of the spinal column and *painfulness upon motion* of the trunk will certainly, even without externally noticeable change in form, count in favor of a developing *vertebral affection*, since even large tumors of the membranes, especially in their favorite location in the dorsal segment, exist, as a rule, without considerable disturbances in movement. Besides this in doubtful cases, the detection of signs of recent or arrested *tuberculosis*, of *fever*, or *gravitation abscesses* and where *a carcinoma is suspected*, the discovery of a *malignant tumor* in other organs, or the existence of a marked amount of cachexia, are important points for diagnostic purposes.

2. Concerning differentiation from a *pachymeningitis cervicalis*, it is to be remembered that this disease is located preferably in the cervical segment, while the majority of tumors are to be found in and opposite the dorsal segment. Furthermore, the disease progresses *far less rapidly* than a tumor of the medulla spinalis and develops preferably in individuals who *formerly* had *syphilis;* in most instances it exists for years and decades until more serious disturbances in cord conduction are developed.

3. The question, whether an *intra-medullary* tumor is present, or one emanating from the meninges can, in many cases, not be answered with certainty. A pronounced *neuralgic prodromal stage* presupposes in the first instance an *extra-medullary* location. For even if the spinal cord tumors may be accompanied by pains, due to concomitant chronic meningitis and to irritation of intra-spinal tracts sensitive to pain, nevertheless it is usually a matter of only transitory and slightly pronounced sensory symptoms of

irritation. But unfortunately, in recent times, observations of meningeal tumors with *painless course* have become more frequent, so that the absence of a neuralgic stage *by no means excludes* an extra-medullary seat. The experience mentioned above, is important, viz., that the neoplasms originating in the cord, especially if a glioma is connected with formation of cavities, often evoke in the beginning of the disease partial sensory paralysis. A further diagnostically important sign of *intra-medullary* tumor consists, as has already been mentioned, in the fact that they not infrequently display a rapid growth upwards and downwards and thereby simulate the picture of a *subacute ascending myelitis*, whereas for the meningeal tumors, it is the slight displacement of the upper boundary of the sensory disturbance, in spite of increase of the symptoms of paralysis, that is characteristic.

4. The syringomyelias may be distinguished from the meningeal tumors mostly by the fact, that a real neuralgic stage is always absent in them, even though slight pains and paræsthesias form no unusual concomitant phenomenon; that furthermore, the sensory disturbance bears a typically *dissociated* character and is confined to a *cutaneous region, supplied by a definite segment of the spinal cord*, while in a pressure paralysis of the cord, by interruption in the conductivity of the sensory tracts, *sensory paralyses of one or both sides* appear below the point of compression. To this must be added the further fact that syringomyelia progresses more quickly upwards, possibly even into the medulla oblongata, and only after a much longer period and in long-continuing stationary intervals gives rise to such severe paralyses of the lower extremities as are usually developed in unbroken succession in meningeal tumors as soon as the first signs of a compression of the cord have appeared.

5. For the differential diagnosis between extra-medullary tumors and that form of a localized serous meningitis, which has been described by *Oppenheim and Krause* and which can imitate in a very deceptive manner the disease picture of a tumor, we lack at present any secure foundation.

Determination of the Location of the Tumor.—The exact determination of the *height* at which the tumor is *located* in the spinal cord is of the greatest practical importance, since upon it, in the first place, depends the success of operative interference.

As an important indication for determining the location, an occasionally demonstrable *locally circumscribed vertebral tenderness* may be serviceable. This will be especially important, if, at the same time, neuralgias appear in a root area, the appertaining spinal-cord segment of which, according to the knowledge we now possess, lies opposite to the vertebra that is sensitive to pressure. *Tenderness to pressure, symptoms of root irritation and of functional loss*, however, must be in harmony with one another, since the sensitiveness of the spinal column as a result of chronic meningitis or for other unknown reasons, is frequently far more extended than would correspond to the size

of the tumor and an isolated area of pain on pressure can occasionally develop on a purely nervous foundation.

In the majority of cases, local tenderness to pressure is not demonstrable, and even pains may be entirely wanting, so that we are limited to the *sensory and motor symptoms of loss of function.*

The possibility of establishing the location of a tumor in a certain segment of the spinal cord, on the basis of sensory phenomena of functional loss, rests upon our knowledge of the *relations of cutaneous sensibility to the individual segments of the spinal cord.* Each of these, namely, receives the majority of the sensory fibres of a definite rather sharply defined cutaneous region. If, therefore, a single segment is injured by the presence of a tumor, a disturbance in sensation makes itself felt, as a rule, in the corresponding region of the skin. As may be seen from the scheme of *Edinger* presented herewith (Fig. 86) the cutaneous regions belonging to the *dorsal segments* of the spinal cord form *stripes that course horizontally upon the skin of the trunk*, so that after the destruction of a dorsal segment a horizontal boundary of sensory disturbances appears upon the trunk.

Clinical observation has taught us, furthermore, that after injury of a single segment, *no complete loss* of sensation, but only a lowering, a *hypæsthesia*, appears in the appertaining stripe upon the skin. This rests on the fact that every individual cutaneous region sends its nerve fibres to *two or three segments placed one above the other*, so that *several of them* must be injured by a tumor, if a *total anæsthesia* is to be found in a definite cutaneous area. This condition, that the lesion of a single segment of the spinal cord causes in the appertaining cutaneous area only a lowering, but not a total loss of sensibility, is of special importance for the establishment of the upper limit of a tumor.

If a tumor injures the spinal cord at a certain level by pressure, there occurs in the place of strongest compression of the cord *an interruption in the conduction of the ascending sensory fibres of the spinal cord.* As a result of this, there is a *general sensory paralysis in that entire part of the body which lies below the point of compression.* The tumor, however, causes also *local segmentary disturbances*, since its upper end, which generally extends above the region of the most severe transverse lesion, affects also the spinal cord segments lying above this place, and gives rise to disturbances in sensibility in their appertaining cutaneous areas.

The scheme of Fig. 86 serves best for the explanation of these relations. By the cross lines is meant that in the lower extremities and in the trunk up to about a hand's breadth of the sternum, the ability of perceiving sensations is *completely lost*, and that *above this region* with total anæsthesia there is a cutaneous stripe (oblique lines, uncrossed), which shows only a *diminution of sensation.*

This *hypæsthetic cutaneous area*, which therefore forms the upper limit of

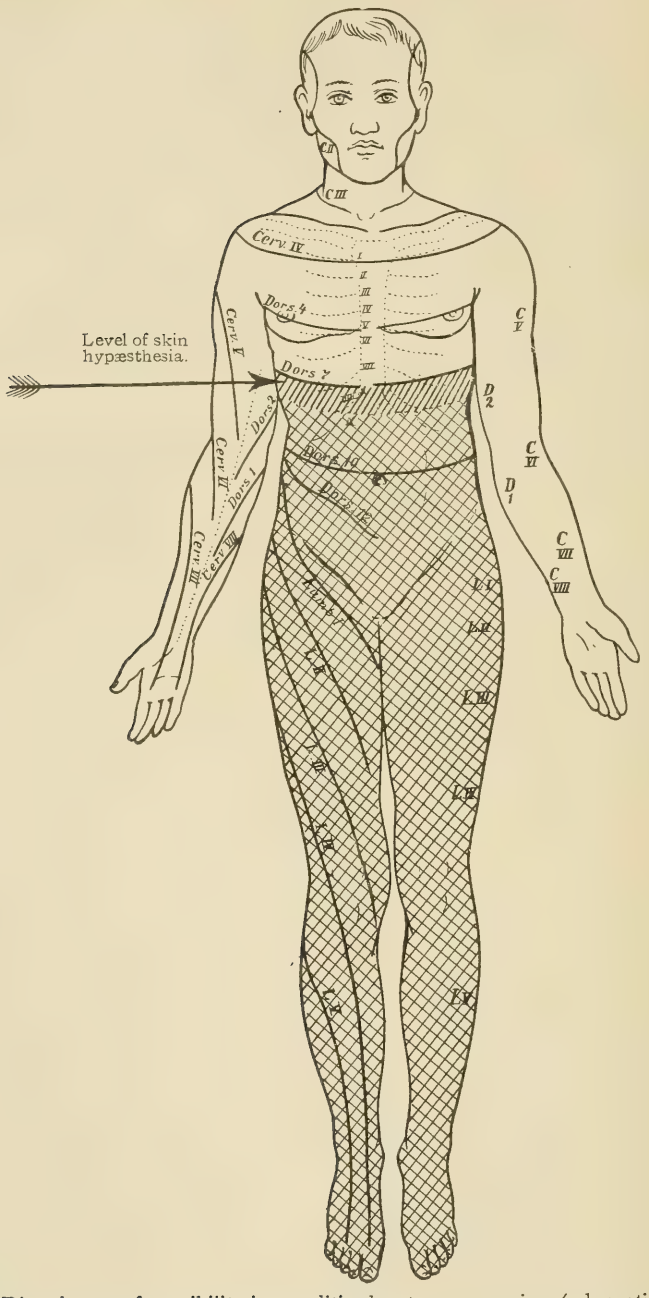


FIG. 86.—Disturbance of sensibility in myelitis due to compression (schematic). The disturbance in sensibility reaches in the trunk the cutaneous region of the 7th dorsal segment. The hypæsthetic cutaneous area is shown by simple cross lines. The double cross lines show the anæsthetic area. (*After Edinger.*)

the sensory disturbance, according to *Edinger's* scheme belongs to the seventh dorsal segment.

According to the above explanation, we must interpret the origin of the sensory paralysis thus schematically presented in this way, that through the pressure of a tumor an interruption in conduction of the sensory spinal-cord tracts lying below the seventh dorsal segment resulted, which caused a total anæsthesia of that part of the body lying below the place of compression. But we must assume in addition, that the upper end of the tumor extends to the seventh dorsal segment, since the cutaneous area belonging to it still exhibits a diminution in sensibility.

If we review the course of the investigation in determining the location of a neoplasm, it is in the first place a matter of the most accurate determination that is possible of the *upper boundary, which shows barely demonstrable sensory disturbances*. With the aid of the well-known sensory area schemes of *Edinger, Seiffer*, and others, we get our bearings to which spinal-cord segment the cutaneous area belongs, which according to the result of the investigation forms the upper limit of the sensory disturbance. If, e. g., the upper limit, as drawn in the figure, reaches to the height of the seventh rib upon the trunk, according to *Edinger's* scheme, the hypæsthetic cutaneous stripe corresponds to the seventh dorsal segment; we may therefore expect the upper end of the tumor to be operated on to be opposite $D7$. As may also be seen from *Gower's* scheme (Fig. 84 on p. 383), $D7$ (oblique lines) lies opposite to the sixth dorsal vertebra and fifth dorsal spinal process. In order to expose the tumor, therefore, the operator will make his incision at the level of the fifth dorsal spinous process and remove the sixth and the seventh, possibly also the fifth vertebral arch, since experience has shown that the seat of the tumor is *generally* assumed *too low* rather than too high. In otherwise correct diagnoses of the tumors in several cases the neoplasm was not found because of a laminectomy that was performed too low. The *longitudinal extent* downwards cannot, as a rule, be determined, especially when the location of the tumor is in the dorsal segment, since this causes in addition to the local phenomena of functional loss and irritation at the upper boundary, general disturbances in conduction below the point of compression due to the pressure on the entire transverse section of the spinal cord, which prevent us from recognizing the segmentary arrangement of the sensory disturbances.

For the determination of the location of the neoplasms in the cervical and lumbar segments, we must consider not only the sensory, but also the *motor* phenomena of functional loss and the *disturbances of the reflexes*. The distribution of the muscular innervation among the various segments of the spinal cord shows the same regularity as the sensory supply of the skin, in as much as *every muscle* is subserved by *several neighboring segments*. Our knowledge, which is as yet incomplete of the segments belonging to the individual muscles is summarized, as it were, in *Edinger's table*. The

localization diagnosis at the levels of the lumbar enlargement and the conus terminalis is particularly difficult, because the roots of this region after their exit from the spinal cord, descend for some distance before they leave the spinal canal. As *Schultze* proved, in a corresponding localization of the tumor in the conus terminalis, the *same symptom complex* may arise as in a *compression in the region of the last lumbar vertebra, or of the sacral vertebræ.* These conditions will be discussed in more detail in a separate chapter on diseases of the conus.

Treatment.—As soon as a suspicion of *spinal cord syphilis* is aroused, or of *tubercular caries*, which in so many cases can not be excluded, our first task is to order *antisyphilitic* treatment, or to try *treatment by extension.* In tumor, after extension treatment of the spinal column, the patient grows worse with unusual frequency; but this has occasionally been observed in caries also, so that no decisive importance can be attributed to this phenomenon. If the treatment fails and the disease progresses, we face the question of *operative interference*, which because of the unusually favorable results that have heretofore been attained in meningo-spinal tumors, must always be taken into consideration.

In every case, in which the *typical symptomatic picture* of a *meningo-spinal tumor* is present, and a *determination* of the *locality seems possible*, surgical interference is earnestly urged for even if a mistake in diagnosis is occasionally unavoidable, since the typical disease picture of an operable meningo-spinal tumor is simulated by vertebral disease or by a tumor of the cord, nevertheless, the chances of complete cure by removal of the usually benign meningo-spinal tumor, are quite favorable.

We are also absolutely justified in suggesting to the patient an *explorative laminectomy*, even though the *atypical* disease picture indicates a *progressive compression* of the spinal cord, and we are unable even after long observation to make a certain diagnosis between intra- and extra-medullary tumors. As long as in the progressive course of the disease, a *meningo-spinal tumor cannot be excluded*, an exploratory opening of the spinal canal is advisable, since without an operation the patient is steadily progressing to certain death. An operation, if the symptoms point to *multiple tumors*, is futile.

Prognosis.—The chances of recovery are naturally most favorable, if the operation is performed *as soon as possible*, before the development of the most severe disturbances in conduction. As a rule, a diagnosis of a tumor and the more exact determination of its location, will be possible only after the development of pronounced phenomena of compression on the part of the cord. But even considerable disturbances in conduction, complete paraplegias and bladder disturbances may *absolutely disappear* in the course of weeks or months, if the pressure lesion has not lasted too long.

The most favorable situation of tumors for an operation, is the dorsal

portion of the spinal cord, less favorable the cervical and lumbar portions, as well as the cauda equina region.

(d) Pachymeningitis Cervicalis Hypertrophica (Charcot)

That form of *chronic inflammation of the membranes of the spinal cord*, described by *Charcot* under this name is a very rare disease. Anatomically the most striking finding is a very considerable *thickening of the dura mater*, which may possibly attain from five to ten times its normal size. The soft membranes likewise undergo inflammatory changes, becoming adherent to each other and to the spinal cord. The microscopic picture shows not only dense, newly formed bands of connective tissue, but also regularly a considerable *nuclear infiltration and a thickening of the vessels* of the spinal cord as well as of its membranes. The inflammatory process does not, in all probability, begin always in the dura, but originates also in the soft membranes; it is usually most marked in the cervical portion; frequently, however, it extends upwards to the meninges of the brain and downwards to the lumbar segment of the spinal cord.

By mechanical compression and extension of the inflammatory process, the roots and the cord itself, the latter at first especially in its marginal portions, become injured. But also more extended degenerations of the nerve fibres with chronic inflammatory changes of the cord and secondary ascending and descending degenerations may develop.

Of its *ætiology* we know only with certainty, that the disease is very frequently preceded by a *syphilitic infection*, while the importance of the other ætiological factors held responsible, like alcoholism, colds, over-exertion, is quite uncertain.

Symptomatology.—The first phenomena of the disease, which may constitute the whole clinical picture for several months, consist of *pains* beginning in the back of the *neck* and *radiating thence* to the *back of the head, the shoulders and arms*. These spontaneously appearing pains, increasing upon pressure and motion, are referred preferably to the region of the ulnar and the median nerves. To these phenomena of irritation, based on compression of the posterior roots, there are added later objective sensory disturbances, likewise *paralysis and atrophy* in the *muscles of the arm and hand*, due to degeneration in the anterior motor roots. Through the fact, that the radial region is frequently spared for some time, a peculiar position of the hand and fingers develops, due to the overbalance of the extensors of the hand and fingers: dorsal flexion of the entire hand and extension of the basal phalanges, while the middle and terminal phalanges are flexed (preacher's hand). During the advance of the process, there are added, sooner or later, to the root symptoms, *signs of a compression of the cord:* slowly increasing paresis of the legs with considerable exaggeration of the

tendon reflexes, mostly slight sensory disturbances in the lower extremities, disturbances in functions in the bladder and rectum.

Course and Prognosis.—The disease, as a rule, extends over many (15 to 20) years. Death may result from decubitus or cystitis. Improvement and standstill are observed in every stage.

Diagnosis.—Differentiation of the disease, is as a rule impossible at the beginning, since the compression paralyses due to meningo-spinal tumors and tumors of the spinal column as well as to tubercular caries, may, at first, evoke similar symptoms of root irritation, without the possibility of always demonstrating the early vertebral changes. Only the course that drags on for years and the simultaneous absence of deformities in the spinal column will point the way to the diagnosis.

Treatment.—Antisyphilitic treatment is of chief importance: iodide of potassium for some time (10 gr. three times a day) and mercurial treatment. As a derivative in more violent pains, the painting of the region of the neck with iodine and ferrum candens may be recommended in particular, in addition to medication with anti-neuralgics.

III. The Syphilitic Diseases of the Spinal Cord

Syphilis as an ætiological factor plays an extraordinarily important rôle in the diseases of the spinal cord. We leave out of consideration, here, such diseases of the spinal cord as in individuals formerly infected with syphilis usually develop slowly, in the form of *primary degenerations* of definite spinal-cord tracts and systems of fibres and are denominated *metasyphilitic* diseases (tabes dorsalis). Here we are concerned only with those disease pictures, in which the pathologic anatomic changes bear a *specifically syphilitic* character, or in which, owing to a series of concomitant conditions, the assumption, that the *diffuse changes in the spinal cord*, which are at the basis of the disease, rest upon syphilis, is made *highly probable*, in spite of the fact that the anatomic signs that characterize the disease are not, in every case, demonstrable. The most frequent and important form of spinal-cord syphilis is

Meningomyelitis Chronica Syphilitica

Anatomical Findings.—The process begins in the soft membranes of the spinal cord, less frequently does it arise from the inner surface of the dura; the membranes are thickened over a large area and permeated in a diffuse manner with a dirty and jelly-like *gummatous granulation-tissue*. Circumscribed gummatous tumors are rare. Beginning from the gummatous new growth, which surrounds the spinal cord like a rind, prong-like cellular proliferations are seen advancing into the marginal portions and extend even

deeper into the gray matter of the cord (Fig. 88); the *roots of the spinal cord* likewise usually exhibit signs of a considerable *infiltration with round cells*. Through the granulation tissue, which enters the substance of the cord like a wedge and through the frequent and usually simultaneously appearing *end-arteritic* changes of the vessels of the spinal cord, the disease is spread to nerve tissue. Due to the *pressure* of the disease foci advancing from the periphery in the form of prongs an *atrophy of the nerve fibres* results. To this is added as further disturbance the *deficient blood supply* of the substance of the spinal cord through the diseased vessels, the thickening of whose walls may bring about a complete closure of the lumen of the vessel; in addition to atrophy of nerve fibres, there are found *ischæmic softenings* of the cord substance, small *hemorrhages* and occasionally even *cavity formations*. The gummatous meningitis in most cases is most strongly developed at the posterior part of the spinal cord. If the process, as has been observed in some cases, remains limited to a marked degree to the roots of the spinal cord, so that many of them appear thickened like knots and swollen, the condition

FIG. 87.—Multiple root neuritis in syphilis. (*After Buttersack.*)

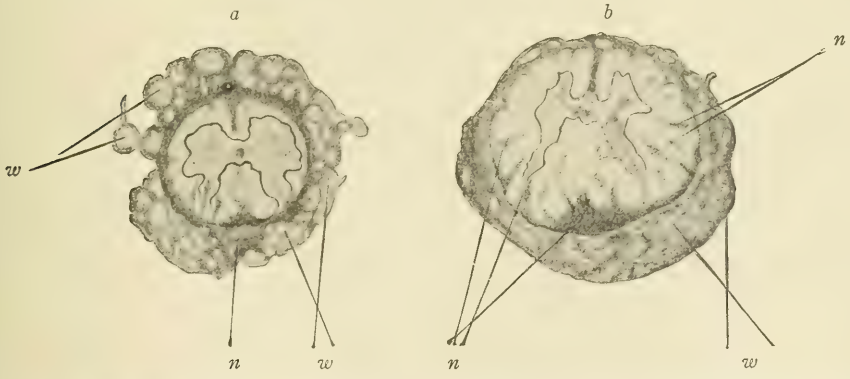


FIG. 88.—Meningomyelitis syphilitica. Penetration of the cord by the new growth. *n*, New growths; *w*, roots of the spinal cord imbedded in the gummatous layers. (*After Boettiger.*)

is spoken of as a *multiple radicular neuritis* (Fig. 87). All the changes here described may be found also in congenital syphilis of the spinal cord.

Symptomatology.—It is easily understood, that with the variety of anatomical changes caused by syphilis, the disease picture may be a very variable one, differing according to the localization and the predominance of lesions of the nerve tissue as caused either by *arteritic* processes, by *gummatous meningitis or by radicular neuritis*. But *Oppenheim* has demonstrated that a symptom complex may be mapped out for the syphilitic form of *meningomyelitis* which, because of the simultaneous appearance of men-

ingeal, *radicular and cord symptoms* as well as through certain *peculiarities of the course*, bears an imprint so characteristic, that the diagnosis in many cases may be made quite easily.

Besides *local pains* in the back and in the sacral region there appear *radiating* pains due to root-irritation, which, according to the location of the disease, are described as "girdle-feeling" or as neuralgias in the extremities. Since first and predominantly the posterior half of the spinal cord is affected, motor-root symptoms appear, as a rule, if at all, only later and then in the form of *partial atrophic paralyses* in the arms and legs. To these meningeal and radicular irritation phenomena are added the important symptoms of a *lesion of the cord:* unilateral or bilateral spastic paresis of the legs caused by the affection of the dorsal portion of the cord. If the cervical portion is affected in its enlargement, the paralysis of the arms is flaccid and atrophic, and is associated with spastic phenomena in the legs, while, if the lesion is located in the lumbar portion, the paralysis—restricted to the lower extremities—is flaccid, from the first. The development of the disturbances in motility may develop *gradually* or quite *suddenly;* likewise, at any time, a slight paraparesis may quickly change into a total paraplegia. Objective disturbances in *sensation* and in the *functions of the bladder and rectum* are found regularly. Quite frequently the picture of a *Brown-Séquard* paralysis appears temporarily as an effect of a more unilateral compression. If, at one of the stages of the development of the disease, the posterior roots are more extensively and more markedly affected by the gummatous changes, a disease picture may occasionally appear showing *absence of the patellar reflex*, *lancinating pains*, sensory disturbances, which closely resembles a *tabes dorsalis*, especially if to the spinal phenomena, there are added cerebral symptoms analogous to those of tabes—*pupillary rigidity*, *paralysis of the muscles of the eye*—as a result of basal meningitis (*Oppenheim*). Such cases have been spoken of as a *pseudotabes syphilitica.*

The special peculiarity, which the course of a meningomyelitis syphilitica is constantly showing, is, that frequently, a very *striking change* in the intensity and extent of the symptoms just described may be observed. A repeated appearance and disappearance, now sudden, now gradual, of single paralytic phenomena is no unusual occurrence. One sees abducens and oculomotor pareses disappear again after existing but a few days, slight parapareses of the legs quickly being transformed into severe paraplegias; pronounced bladder disturbances may disappear again in a short time; in like manner the neuralgic troubles caused by radicular irritation show considerable fluctuations in severity and extent. *Oppenheim* has called attention in particular also to the *condition of the tendon reflexes*, which may appear diminished, abolished, or actively increased, all within a few days. It has been attempted to explain this change in individual symptoms, which is occasionally sudden and always characteristic of lues spinalis by the fact

that, as a result of the change in the amount of blood the tissues, the granulation tissue, the roots of the spinal cord and the cord are exposed to pressure of varying force. Another important sign of lues spinalis depends upon the fact that the syphilitic process does not, as a rule, confine itself to the spinal cord, but attacks also the *meninges* and the *vessels of the brain* and above all, *basal cerebral phenomena* appear at an early period. In the majority of cases, therefore, there will appear, in addition to the spinal symptoms, also some few cerebral paralytic symptoms—disturbances in the muscles of the eye or in vision—which exhibit the same instability as do those of the spinal cord.

In the year 1892 Erb separated another disease entity, under the designation of *syphilitic spinal paralysis*, from the large and varied group of affections of the spinal cord, which are based upon syphilis. He had suspected, that similar anatomic changes would be found at the basis of the symptom complex sketched by him, as those of the meningomyelitis just discussed, that is, in particular, specific myelitis, or arteritic and gummatous processes. This hypothesis has not found support in the postmortem findings that are available thus far. For it has been shown that various changes in the spinal cord come under consideration as the pathologic-anatomical foundation of *Erb's* symptomatic picture. In a number of cases, a sclerosis of certain tracts of the spinal cord, of the pyramidal and lateral cerebellar tracts and of the columns of *Goll* were found, without any participation of the meninges, or any specific changes in the vessels being noted, so that anatomically, the picture of a *primary combined system disease* was present. In other cases, however, a chronic, disseminated or partial *myelitis transversa* was found with ascending and descending secondary degenerations, or even a combination of *diffuse chronic myelitis* with apparently primary columnar degenerations. The thickening of the walls of the vessels, which was demonstrated especially in the chronic myelitic foci, did not usually exhibit the picture of specific end-arteritic changes, even though some observers, like *Nonne*, proved the presence of apparently *specific end-arteritis* in some arteries, in addition to these ordinary thickenings in the walls of the vessels.

Erb's disease picture, in its *pure* form, is rather clearly defined. In individuals, who previously have been infected with syphilis, there develops gradually, and usually in an insidious manner, more commonly in the earlier stages of syphilis, about two to six years after the infection, a *spastic paresis* of the legs with considerable *increase of the tendon reflexes* and the Babinski reflex. The gait is distinctly spastic, even if the muscular rigidity is but slightly pronounced. Besides the weakness in the lower limbs, there appears as a regular and early symptom, which sometimes precedes all the other phenomena, a *disturbance of the bladder*. The *sensory disturbances and paræsthesias*, which are demonstrable with equal frequency, are usually *of slight degree*. Variations from this symptomatic picture may

occur as follows: the beginning of the disease may occasionally be rather acute with quickly developing paraplegia and muscular contractures. In the pure form of syphilitic spinal paralysis, according to Erb, we always note the absence of *more severe pains*, of *more severe sensory disturbances*, as well as of all phenomena which point to a *disease of the brain or its membranes*.

The course of the disease is, as a rule, *slow*, extending over *many years*, with occasional improvements and standstills. But there is also a form with a more rapid course and a fatal termination after a few years.

Diagnosis.—Considering the frequency of spinal diseases originating upon a syphilitic foundation, and the ability of syphilis to appear through its *multiple distribution* in all the different parts of the nervous system in the most varied symptomatic pictures, it would be well always to think of syphilis in all diffuse diseases of the spinal cord, unless we have before us absolutely typical disease pictures.

In our inquiry into the previous history and our investigation of all the other organs of the body, we must make it our chief aim to look up former diseases, in which there was a suspicion of syphilis, and notice with special care all signs of *recent* or *past lues*. According to experience, however, our endeavors in this direction are not rarely entirely fruitless, even where the disease of the spinal cord proves later to be surely syphilitic.

Differentiation of a *meningomyelitis syphilitica* from a simple acute or subacute *transverse myelitis* and from a *compression paralysis* of the spinal cord, will as a rule, cause no difficulties. The more severe pains and a neuralgic prodromal stage do not belong to the disease picture of an acute myelitis. As contrasted with a caries, which exists for a long time without deformity of the spinal column, sufficient signs for distinction will be found in the fact that the compression paralysis, as a rule, has a gradual *steady* course, whereas the lues spinalis is characterized particularly by the coming and going of phenomena and usually creates a *disease picture pointing to multiple foci*. The distinction from *multiple sclerosis*, which likewise frequently exhibits a striking change of sudden deteriorations and improvements, can occasionally cause difficulties. The differential diagnosis will mostly be based on the fact, that pronounced *sensory phenomena of irritation* are absent in sclerosis and that, on the other hand, the characteristic *intention tremor, the speech disturbance*, the partial *optic atrophy* and *nystagmus* are foreign to lues spinalis.

Both *multiple sarcomatosis and carcinomatosis of the membranes* may give rise to a confusion with lues spinalis. Here only the further course and the result of specific treatment will make diagnosis possible.

Erb's syphilitic spinal paralysis, in its pure form, presents an easily defined disease picture; it is distinguished from the simple *spastic spinal paralysis (Erb)* by the presence of sensory and bladder disturbances, from meningomyelitis by the absence of sensory meningeal and radicular irritation symp-

toms and the absence of cerebral phenomena. In differentiation from combined disease of the posterior and lateral columns, the course of the disease must be specially considered. In combined columnar disease, there is a steadily progressing process, which step by step, attacks one system after the other. The course of Erb's disease is not, on the other hand, so regular; *standstills and improvements* are not unusual; likewise even total *disappearance* of single symptoms under specific treatment.

Erb himself emphasized the fact, that a large number of transitional forms with their *deviations*, which are, in part, considerable, *from the pure picture of syphilitic spinal paralysis*, may often exhibit far-reaching resemblance to the most varied forms of lues spinalis, so that certain differentiation, particularly from meningomyelitis, is frequently impossible. Many prominent neurologists advocate the point of view (*Oppenheim* and others) that the symptom complex described by Erb represents only an *initial stage*, or the *picture of one phase* of meningomyelitis chronica syphilitica.

Prognosis.—Complete *recovery* from lues spinalis is possible; it is to be expected particularly when the disease is treated early, before the phenomena of a lesion of the cord have appeared. But even after the development of an incomplete transverse lesion, there is still, according to *Oppenheim's* experience, possibility of absolute restoration. In the great majority of cases, however, the outcome of the disease, as soon as the cord itself has suffered, is quoad restitutionem, unfavorable, as more or less considerable functional disturbances remain in the form of spastic pareses, bladder disturbances, etc. The prognosis even in the cases whose course is favorable is, nevertheless, made less favorable by the fact that a relapse may occur at any time.

Treatment.—The treatment consists in inunction which is continued for several weeks (using 1–2 drams ung. hydrarg.) with simultaneous administration of iodide of potassium (at the beginning 15 grs.; later, 60 grs. daily). *Oppenheim* recommends a rapid increase in the dosage of iodide of potassium, or to give large doses at once (up to 200 and 300 grs. pro die); he has also had results from internal or subcutaneous employment of iodipin (10–20 g. of the 25% solution injected daily for 8–10 days), when mercury and iodide of potassium remained ineffective.

The inunctions of 1 to 2 drams *ung. hydrarg.* pro die, had better be made in the evening before going to bed, since the absorption of the mercury is effected in the inunction treatment, mainly through *inhalation of the mercurial fumes*. The most suitable places upon the body to be used alternately, have been shown to be the *calves, the outer and posterior surface of the thighs, the region of the abdomen and the loins, the lateral parts of the thorax, the outer and posterior surface of the fore- and upper arm.* According to experience, the following parts have proved sensitive to the salve, the flexor aspect of

the joints, the axilla, the inner surface of the thighs and all places upon the skin with but a small amount of underlying softer parts as, e. g., the skin over the tibias. Between each group of five days, on which inunctions but no baths are given, one sandwiches in a sixth day, in which the patient is treated only to a warm cleansing bath. To avoid a mercurial disturbance of the mucous membranes of the mouth, which in its initial stage, manifests itself by an increase in the secretion of saliva, a metallic taste in the mouth and a feeling as if the teeth are growing longer, persistent care of the mouth is essential. The mouth should be cleansed *after every meal.*

A repetition of the inunction treatment is necessary, as soon as new symptoms of disease appear. The question, how long shall the treatment be continued, *Gowers* answers by stating that after a course of treatment extending over 6 to 10 weeks without visible results, further continuation for the time at least is useless. If there has been improvement in the disease, and a standstill has been reached, it seems advisable to order *regularly* in the next few years, one or two courses of antisyphilitic treatment, even if relapses have not occurred. The combination of the inunction treatment with baths in various resorts (*Aachen, Toelz, Nenndorf*) is frequently recommended. In addition to the specific treatment, it is important above all to regulate the patient's entire manner of life: avoidance of all *physical and mental overe-xertion* and of all *sexual* and *alcoholic* excesses.

IV. Myelitis

Definition.—We are not at present in a position to draw a sufficiently sharp line from the clinical or pathologic-anatomical point of view between disease pictures, which are based on acute and subacute changes in the spinal cord with the signs regarded as characteristic of an inflammation, and those, in which *phenomena* of acute *degeneration and disintegration* of the cord substance are preponderantly demonstrable. Most clinicians, therefore, classify for the present as myelitis all *diffuse and disseminated inflammatory* and *softening processes* in the spinal cord, in so far as they are not the result of a direct crushing or contusion of the cord.

Classification of the Forms of Myelitis and Anatomic Findings

One distinguishes a *transverse myelitis*, in which a focus, variable as to the height of its location, takes up all or the greater part of the transverse section of the spinal cord; also, a *myelitis disseminata*, with numerous smaller and larger foci strewn over the entire length of the medulla spinalis, usually associated with participation of the medulla oblongata and the brain; and finally, *poliomyelitis*, which depends upon inflammatory changes, localized mainly in the anterior gray horns.

The more recent investigations have shown that a separation of these different forms of myelitis is justified only in so far as by the nomenclature attention is called to the chief focus of the disease and the special localization of the changes through which the disease picture attains its peculiar stamp. As a rule, both in acute *myelitis transversa* and in *poliomyelitis*, there are, in addition to the main focus, countless small and minute inflammatory foci, scattered over the gray and white matter, so that in both these forms therefore, there is exhibited the anatomic picture of a *disseminated myelitis*.

A diffuse myelitic focus may be recognized even by palpation and by macroscopic inspection, because of its softer consistency and the more vivid injection of the vessels of the soft membranes in contrast to the adjacent sound parts of the spinal cord. When the softening is very pronounced, the grayish-yellow discolored substance of the cord flows over the surface of an incision, while in less marked development of the changes, only the lines of separation between the gray and white matter seem blurred. The microscopic appearance of the hardened preparation may vary considerably. Occasionally the inflammatory changes of the vessels are comparatively unimportant as compared with the acute phenomena of degeneration of the nerve elements. The *axis cylinders and medullated sheaths* seem to be much *swollen* and *inflated like bubbles*, either singly, or in groups, so that the volume of a nerve fibre may often amount to from 10 to 20 times its normal size. Through disintegration of the diseased fibres, there are formed *spaces* in the tissue between the nerve fibres, which are still unchanged; these spaces are filled with granular detritus, cord remnants and drops of fat (Fig. 89). As the glia tissue also, at first by swelling, and later by disintegration, participates in the process, softening foci develop. Compensation takes place through the proliferation of the interstitial tissue in the neighborhood of the focus of degeneration; a sclerotic scar tissue is formed. During this process, the entire focus contains numerous, so-called granular cells, which are loaded with disintegrated particles of tissue and are to be found especially in the perivascular spaces of the vessels, which are usually thickened.

In some cases, especially in myelitis disseminata and in poliomyelitis, it is not only the phenomena of acute degeneration in the nerve parenchyma, which occupy a prominent position in the anatomic picture, but also *inflammatory changes in the vessels*. They are present in the form of pronounced dilatation of the arterial and venous pathways, possibly with an extravasation of blood into their surroundings and dense accumulation of round cells in the adventitia of the vessels and the perivascular lymphatic spaces. After all more extensive processes of inflammation and softening in the spinal cord, there is a development of "*secondary degenerations*" of the ascending and descending spinal-cord tracts, which have become separated from their

ganglion cells. The soft *membranes of the spinal cord* are, as a rule, also affected in myelitis. Even in acute poliomyelitis, in recent cases, changes in the pia were found by *Dauber*, *Schultze*, and others in addition to the changes in the vessels.

Bacterial Findings.—The presence of bacteria in the inflammatory foci, the meninges and the cerebro-spinal fluid, has been demonstrated, heretofore, only in rare cases, even when the disease appeared in direct connection with infectious processes. In the diseased tissue of the spinal cord itself

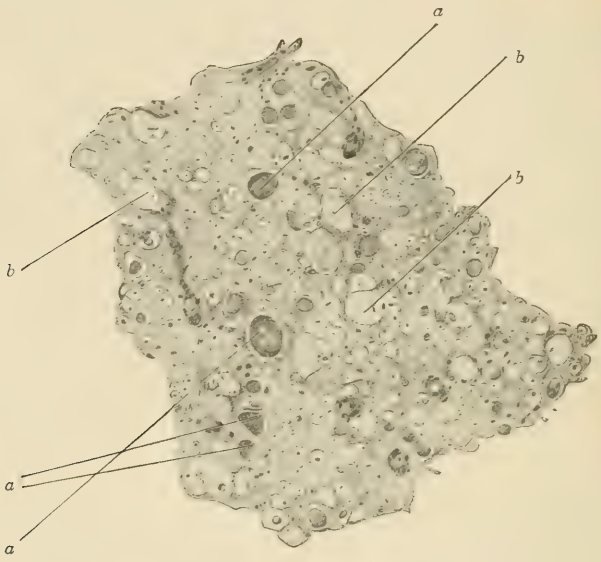


FIG. 89.—Myelitic focus in acute myelitis with numerous swollen axis cylinders (*a*) and lacuna through disintegration of nerve fibres (*b*).

have been found in addition to *tubercle bacilli*, *strepto-staphylo- and pneumococci*, *also the bacilli of anthrax and typhoid;* in the cerebro-spinal fluid, in poliomyelitis, the presence of extra-cellular *diplococci* was demonstrated by *Schultze* and others, the more exact determination of which as meningococci by cultural proceedings was not possible; *Strümpell* was able to isolate staphylococci in the fluid of a patient with myelitis, which had supervened in a case of whitlow.

According to the experiments of *Hoche*, *Homen* and others, the rarity of bacterial findings may be due to the fact, that the bacteria after their penetration into the substance of the spinal cord and into the cerebro-spinal fluid, quickly disappear again.

Ætiology.—In the ætiology of acute myelitis, in all its different forms, *infections* and *intoxications* play the most important rôle. There is probably no acute infectious disease, in the wake of which the appearance of a myelitis has not been observed. I must mention especially *smallpox*, *typhoid*,

diphtheria, *influenza*, *erysipelas*, *measles*, *scarlet fever*, *malaria*, *gonorrhea*. Even in connection with purulent processes in the various organs of the body: in *whitlow*, *perityphlitis*, *suppuration in the antrum of Highmore*, an acute myelitis is occasionally observed. Furthermore, in *syphilis* and *tuberculosis*, acute myelitis occurs, which, anatomically, does not present the specific changes characteristic of these diseases. For the myelitis, which occasionally appears in *pregnancy* and in the *puerperium*, an infectious cause is also assumed. Since investigators have succeeded experimentally, by the introduction of toxic bacterial products, in producing inflammatory changes of the spinal cord, which bear great similarity to those in acute myelitis, it is very probable that myelitis is caused not only by the *local effects* of the bacteria themselves, but also by the toxins produced by them; they may reach the spinal cord by means of the blood and the lymphatics.

Catching cold, *over-exertion*, *and psychic excitement* were formerly thought by many to be the only sources of certain cases of acute myelitis; this seems doubtful in the light of more recent experience; however, one must assign to them the importance of an *influential assisting and exciting cause* under certain circumstances.

After poisonings with chemical substances of the most different varieties —CO, illuminating gas, bisulphide of carbon, chloroform, lead—a larger number of cases of myelitis having a more chronic and subacute course rather than taking an acute form are observed; but acute cases do occur and especially after poisoning by smoke in firemen, suddenly appearing phenomena of spinal paralysis are not at all uncommon.

Those diffuse changes in the spinal cord, which with numerous small *acute foci of degeneration*, develop upon the basis of a *cancerous cachexia in carcinoma*, furthermore, in *pernicious anæmia*, *leucæmia*, *chronic nephritis*, *diabetes* and *gout*, exhibit *clinically*, as a rule, also a more *subacute* or *chronic* character. In some of these cases morbid products of metabolism may be considered to be the cause of the inflammation; in others it may be a question of necroses and softenings resulting from the anæmia and general cachexia, which so frequently accompany these diseases.

A peculiar form of myelitis, observed in *workers* in *tunnels*, and designated *Caisson paralysis*, needs special mention. As a result of a sudden decrease in high atmospheric pressure, there is a development of gas bubbles in the blood, and therewith gas embolism occurs in the blood-vessels including the vessels of the spinal cord. Through these macroscopically and microscopically recognizable embolizing gas bubbles there arise numerous small ischæmic softening foci. Finally, we must mention the rare occurrence of softening as the consequence of an embolic closing of the aorta abdominalis, also embolic and thrombotic occlusions of the arteries of the spinal cord.

Symptomatology.—The phenomena of the disease set in very quickly

in most cases of acute myelitis, so that after a brief prodromal period, during which *slight pains* in the *back and sacral region*, *weariness* and *paræsthesias* in the legs are complained of, the symptoms of paralysis develop more or less suddenly in a few hours or in the course of several days. The onset is usually with *fever;* fever that lasts for weeks and occasional exacerbations of fever with aggravation of the spinal symptoms frequently occur.

Even at the acme of the disease, the inflammatory focus does not, as a rule, extend symmetrically over the entire transverse section, but leaves larger or smaller areas of the gray and the white matter untouched. Accordingly, the grouping of symptoms and the intensity of the phenomena of the paralysis, are dependent, not only on the *height of the localization*, but also and to a great degree on the manner of the diffusion to the individual tracts of the spinal cord. In discussing the spinal phenomena, we shall start with the *most frequent form of myelitis dorsalis.*

1. The disturbances of *motility* stand in the foreground of the disease picture. A *total paraplegia* of the lower extremities develops if the lateral pyramidal and anterior columnar tracts are destroyed by the great extent of the myelitic focus. In an incomplete transverse myelitis, a series of muscles will still be able to carry out their functions, or all movements of the legs will still be possible, but with decreased strength. The paralysis is at first *flaccid;* but very soon there appears a *muscular stiffness*, a *spastic condition*, which can lead first to a position of extension, later to severe *flexion contractures* in the paralyzed legs. Not only in the beginning, but also in the further course of the disease, motor *irritation phenomena*, twitchings, appear in the paralyzed limbs, which partly, at least, depend on the increased reflex excitability, since even slight irritations of the skin, such as moving the bed cover, change of position, etc., suffice to evoke painful muscular contractions; partly, also, they may be the result of a *direct irritation* of motor fibres of the spinal cord. The participation of the abdominal muscles is recognizable even in its slight degrees by the fact that the patient is not able to rise from a supine position, without aiding himself with his hands.

2. The *tendon reflexes* in the legs are usually *increased.* The morbid increase manifests itself in that when the knee-cap or the tibia of the extended leg is lightly tapped, a contraction of the quadriceps muscle results, and in that *patellar and ankle clonus* may be excited. A permanent loss of the tendon phenomena has been observed only in those cases in which the disease approximated a total transverse interruption of the cord. It is noteworthy, that as a result of high degrees of contractures, the excitation of the tendon reflexes may be made difficult or impossible.

The *cutaneous reflexes* are usually retained and actively increased, so that by pinching a fold of the skin in the paralyzed and anæsthetic regions muscular contractions of that or even of the opposite extremity result. The plantar reflex shows the alteration so frequently characteristic of a spastic

paralysis, *dorsal flexion of the great toe* when the sole of the foot is stroked, especially on its outer side (*Babinski's sign*). In more extensive transverse lesions the cutaneous reflexes like the tendon reflexes may disappear also. The abdominal reflexes are partly or entirely lost, if the reflex arc subserved by the posterior roots of the ninth to the twelfth intercostal nerves is destroyed by the location of the myelitic focus in the lower dorsal cord.

3. The *sensibility*, as a rule, exhibits considerable disturbance, even to the total loss of all qualities of sensation in the legs and trunk, up to a level corresponding to the region subserved by the segment of the spinal cord which is affected by the focus. If the process is restricted to a smaller part of the cross-section, the disturbance in sensation extends only to one leg, or parts of the trunk, or consists only in disturbances of certain qualities of sensation, whereby the isolated diminution in *muscle, tactile* and *pressure sense* points to a predominant disturbance in the posterior columns, impairment of the *temperature and pain sensations*, to an affection of the posterior horns.

4. *Bladder disturbances* are frequently an *early symptom* of acute myelitis, which may be discovered even in the prodromal stage. A participation of the bladder and rectum is never absent in any of the more extensive transverse lesions. In the beginning, there is a slight *difficulty in micturition*, which as a result of the paralysis of the detrusor muscle quickly increases to a total *retentio urinæ*. Overdistension of the bladder is occasionally not perceived by the patients at all, as, because of the lesion of the sensory tracts, the conduction of the irritation impulses arising from the mucous membrane of the bladder and going to the cerebrum no longer takes place. As further consequence of the excessive filling and dilatation of the bladder, there appears a slow dribbling of the urine, an ischuria paradoxa. As a rule, after a certain time, the mechanism of the bladder works independently, in such a way that at comparatively regular periods, as soon as the bladder has reached a certain degree of fulness, *reflex evacuation* takes place. The disturbance in rectal activity shows itself usually as a high degree of sluggishness of the bowels.

5. The *trophic disturbances* of the skin are of special importance. Far more quickly than with other bed-ridden patients is an acute *appearance of decubitus* observed in myelitis. As early as the first 24 to 48 hours after the appearance of the severe phenomena of paralysis, bed-sores due to pressure may develop, so that careful treatment of the skin, belongs to the most important duties of the physician, from the very beginning of the disease. The favorite places for bed-sores are, beside the sacral bones, the region of the trochanters, the heels and the shoulder-blades, and the inner surfaces of the knees where they rub against each other. Frequently, also, edema and sclerodermoid conditions develop during the longer continuation of the disease.

6. Of cerebral complications of a focal myelitis only *neuritis optica* is worth mentioning. Other disturbances in the realm of the cerebral nerves are usually absent in the non-disseminated form.

If the seat of the focus is not, as is usual, in the dorsal portion of the spinal cord, but in the *cervical or lumbar portion*, we must expect certain deviations from the disease picture just described.

Myelitis cervicalis is the rarest and most dangerous form of myelitis. If the focus lies *above* the cervical enlargement there is found, besides spastic paralysis of the legs and the usual phenomena of a dorsal myelitis, also a *spastic paresis of the arms;* a *bilateral phrenicus paralysis* is followed by death through suffocation. If, however, the focus is in the region of the *cervical enlargement*, there develops—because of the destruction of the ganglion cells of the anterior horn—a *flaccid, atrophic* paralysis of the arms with electric reaction of degeneration. The synchronous appearance of *oculo-pupillary* symptoms due to sympathetic paralysis—contraction of the pupils and of the palpebral fissures—indicates that the eighth cervical and first dorsal segments are also affected.

In a *myelitis lumbalis*, the upper extremities are quite free; there exists a *flaccid degenerative* paralysis of the legs with *weakening or absence of the tendon reflexes.* The paralysis of sensation is restricted to the lower extremities alone; bladder and rectum are paralyzed.

If the *lower sacral cord* is principally affected, it is preeminently the sciatic region which is paralyzed—in the motor aspect—and the sensory disturbances extend only to the back of the thighs and their inner half to the lower third on the buttocks and the genital organs. Besides, there exist disturbances in bladder and rectum.

Myelitis disseminata requires special discussion, since the disease picture may exhibit many different forms, especially when greater participation of the *medulla oblongata and the brain* is shown to exist (Fig. 90). When through numerous smaller foci a larger part of the transverse section of the spinal cord is deprived of its function, the disease may present clinically so completely the picture of an acute *transverse myelitis*, as to make it well-nigh impossible during lifetime for a diagnosis of disseminated myelitis to be made. Furthermore, a symptom complex may develop which bears the greatest similarity to a *multiple sclerosis* running an *acute* course. In other cases, an *acute ataxia*, associated with loss of tendon reflexes and bladder disturbances, or even with spastic pareses, occupy the foreground of the symptomatic picture. Occasionally, also, it may happen that in the beginning of the disease, the cerebral phenomena are very much pronounced as compared with the symptoms of spinal paralysis, so that the picture of a pure *encephalitis* may be believed to exist.

Course.—As a rule, myelitis reaches the acme of its development within a few days and weeks; death may occur rapidly or possibly an improvement

may ensue (with occasional sharp relapses, which are accompanied by renewed fever), which may lead to complete *recovery*. This, however, is *rare* indeed; usually the severe phenomena of paralysis are but partially recovered from, and there comes to be a permanent condition of illness, in the course of which, death may appear at any time, as a result of decubitus or cystitis.

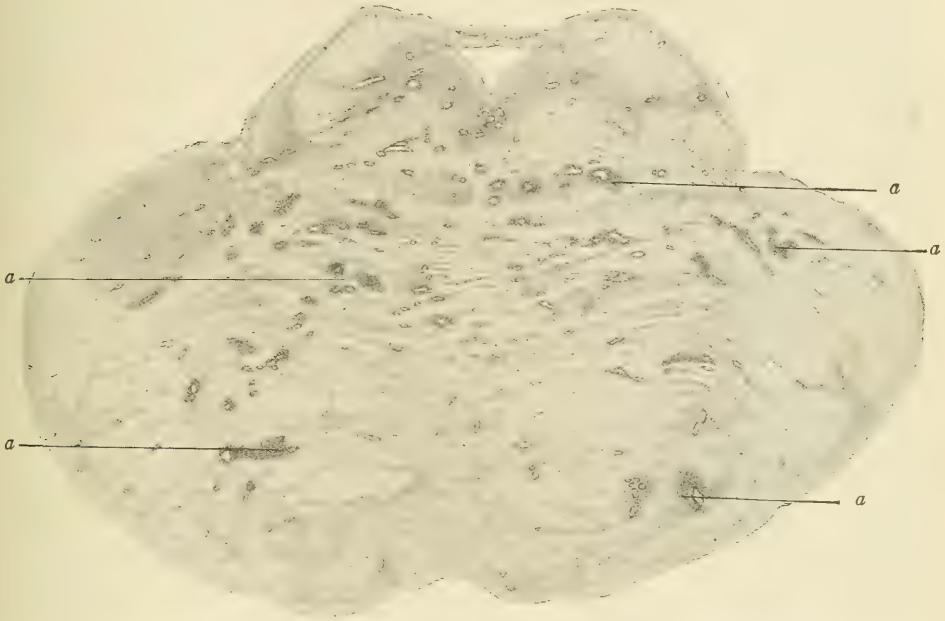


FIG. 90.—Myelitis disseminata. Cross-section through the pons with numerous small inflammatory foci grouped around the vessels (*a*). (*Author's own observation.*)

Chronic Myelitis

in which the disease picture of a progressive diffuse transverse myelitis develops *insidiously* from the first, is an extremely rare disease. Only in isolated cases, heretofore, could be demonstrated an *anatomic* basis for a disease of the spinal cord, exhibiting the picture of chronic myelitis, the presence of a *diffuse chronic myelitis*. In the great majority of cases, another disease is hidden behind the symptom complex of a chronic transverse myelitis, a *multiple sclerosis*, a *lues spinalis*, a *combined column disease*, or a *compression paralysis* of the spinal cord.

In *chronic dorsal myelitis*, the initial symptoms are first of all a slight tendency to *fatigue* in walking with increasing *stiffness in the muscles* and abnormal sensations, *paræsthesias*, in the legs. Actual pains are only transitory, and usually present only in slight degree. The *tendon reflexes* frequently show a considerable *increase* with or without accompanying muscular rigidity. The objective sensory disturbances usually remain slight,

as compared with the *gradually appearing phenomena of motor paralysis.* Bladder and rectal disturbances and impotence are regular concomitant phenomena in advanced stages of the disease. The duration, as a rule, extends over years; there may be a standstill, and long periods of improvement.

Diagnosis.—For the diagnosis of acute myelitis transversa, it will frequently be of importance to show that the beginning of the disease was connected with certain diseases, infections, etc., which, as we know from experience, play an ætiological rôle in acute myelitis. Confusion with suddenly occurring hemorrhage of the spinal cord is possible, since in this disease, also, prodromal phenomena, paræsthesias, twitchings may appear, caused by disturbances in circulation, as a result of diseases of the blood-vessels. Differentiation from a pressure paralysis in caries or vertebral carcinoma, both of which may occasionally develop rapidly, will, as a rule, cause no difficulty, since the *neuralgic prodromal stage* and eventually, *deformities of the spinal column* afford sufficient points for diagnostic differentiation. *Schultze* observed in one instance that a tumor of the spinal meninges (possibly by a transitory damming back of the cerebro-spinal fluid) might rather suddenly exhibit the phenomena of transverse lesion, and thereby simulate an acute myelitis. In such cases, only further observation, the proof of the steady and irresistible progress of all spinal symptoms will make diagnosis possible. Separation from a *polyneuritis*, which may also appear in connection with infectious diseases, will be called into question only when, in *lumbar myelitis*, the ganglion cells of the anterior horns are destroyed, and atrophic degenerative muscular paralyses are present. The *neuritic pains* on the one hand, and on the other the bladder disturbances, which are very rarely found in polyneuritis, are points for differentiation between them.

Confusion with hysteric paralyses is possible especially when, associated with the usual hysteric phenomena, there is present a real transverse myelitis. The *Babinski sign* has proved to be a sure sign for distinguishing between functional and organic paralysis. Beside this, the change in the severity of the phenomena of paralysis, the occasionally striking contrast between the ability to walk and the mobility of the legs when the patient is lying on his back, as well as the effect of suggestive influences, make possible the detection of the hysteric nature of the disease.

In the *differential diagnosis of chronic myelitis*, the most important diseases are compression paralyses due to caries, vertebral tumors and tumors of the spinal cord, sclerosis multiplex, lues cerebrospinalis and combined column diseases. We would refer to the chapters concerning them.

Prognosis.—This is always *serious* in myelitis. The chances of preserving life and of complete or partial functional recovery are the less favorable, the more quickly the phenomena of a total transverse lesion are developed. Especially dangerous is the disease of the upper cervical cord, because of the possibility of the occurrence of *paralysis of the diaphragm.*

Even after the disappearance of the acute phenomena, if there is a stationary condition with partial functional restoration, the disease may at any time terminate fatally in consequence of *decubitus* and *cystitis* with their resulting phenomena. Those cases of myelitis which appear after acute infectious diseases offer the best chance of recovery. *Syphilitic* myelitis which develops soon after the infection, frequently runs quite a rapid course.

Treatment.—In the acute stage our chief aim will have to be the *prevention of decubitus and cystitis*. The early use of water or air beds and frequent bathing with alcohol and water of those parts of the body which are especially exposed to pressure, act as prophylactics with respect to pressure bed-sores which so frequently develop with considerable acuteness precisely in myelitis. For the rest, *absolute quiet in bed* at the beginning of the disease, and even a long time after the disappearance of the symptoms of the spinal paralysis, is necessary. In the *post-infectious* forms, *diaphoretic* procedures and the administration of *preparations of salicylates* are recommended. If there be a *suspicion of syphilis*, iodide of potassium (gr. x t.i.d.) should be tried; mercurial treatment (℥j ung. hydrarg.), in which there is always a possibility that the Hg. may have an injurious effect upon the more severely diseased tissue, must be stopped at once if an unfavorable result is perceived. In previous *malarial infection, quinine and arsenic* may have favorable results. After lumbar punctures we have observed frequently an objective improvement, return of the reflexes, increase in mobility.

If frequent catheterization is necessary, besides very painstaking antisepsis, a prophylactic administration of urotropin (gr. v t.i.d.) is advisable.

In the *stationary* stage with residuary phenomena of pareses and slight spastic conditions, *treatment by baths* is to be tried first. There is need of serious warning against hot baths. Simple warm baths (92°–96° F. and lasting 15 minutes) are most useful, which in case of more severe spastic phenomena may be extended over a long time (1–2 hours). Treatment at thermal resorts where the hot springs contain carbonic acid, *Oeynhausen, Nauheim*, and others like *Ragaz, Teplitz, Wildbad* are to be recommended.

As a means of comfort for the patients with stationary conditions of paralysis, the *galvanic* current is recommended for direct treatment of the spinal cord (3 to 5 minutes of not too strong, stabile galvanic current on the spinal column) and for muscular nerve stimulation in the paralyzed extremities.

We may expect from electricity an effect that will further recovery, only when the phenomena of paralysis are themselves on the point of retrogression. If a bladder disturbance remains, galvanic treatment may be tried, either both electrodes over the lumbar cord, or anode on the lumbar region, cathode above the symphysis or upon the perineum. The strength of the current, in a duration of 5 to 10 minutes, is to be 3 to 5 M. A.

Supplement

Spinal-cord abscess, an extraordinarily rare disease, develops either after an injury to the spinal cord, or is due to metastasis. The formation of an abscess has been observed after *putrid bronchitis*, *prostatic suppuration*, *gonorrhœa*, *malignant endocarditis*, *appendicitis*. Spinal-cord abscess is usually of central location and may become very extensive; usually it is connected with purulent meningitis, which occasionally appears only later, so that *lumbar puncture* may give, at first, a *negative result.*

The disease picture, which is constructed from the phenomena of a diffuse transverse disease with an acute onset, affords nothing characteristic. Usually it will be concealed behind the symptomatic picture of an *acute myelitis* and only by the detection of other *purulent foci*, synchronous general *pyæmic phenomena* and a *purulent exudate* found by lumbar puncture, is the diagnosis made possible.

(7). DISEASES OF THE CONUS TERMINALIS AND THE CAUDA EQUINA

BY

L. R. MÜLLER (Augsburg)

The lowest section of the spinal cord and the nerve roots going out from it, are anatomically so different from the other parts of the spinal cord, that the diseases there have a peculiar character.

The sacral cord with the conus terminalis is, for the most part, situated behind the body of the first lumbar vertebra and extends downwards only to the middle of the second lumbar vertebra. After opening of the spinal canal and of the dura mater the lowest division of the spinal cord is not, itself, visible. From the lumbar enlargement, spring in continuous series thick bundles of nerves, which all running downwards, soon become so numerous that they entirely conceal the lowest part of the sacral cord, the conus. A separation between the sacral and lumbar cord, such as is easily to be seen between the cervical and dorsal portions, or the dorsal and lumbar portions, is to be determined only with difficulty. Only when the bundles of the cauda equina are traced to their exit from the dural sac and from the spinal canal, that is 10 to 14 cm. downwards, can the artificial boundary between the lumbar and sacral cord be determined by counting the separate roots. But even if the boundary between the lumbar and sacral cord can not be recognized morphologically, theoretically, at least, a distinction must be maintained between the nuclear region of the plexus lumbalis and that of the plexus sacralis or ischiadicus. In its lower half, the sacral cord tapers off quickly; that part of the spinal cord which embraces the third, fourth and fifth sacral segment and both coccygeal

segments, is called *conus medullaris*. In this lowest division of the spinal cord, the relation between the anterior and the posterior roots is not so uniform as in the rest of the spinal cord. If more than double the number of posterior roots enter the cord here, as anterior roots spring from it, this must be explained by the fact that no large group of muscles are to be innervated from the third sacral segment downward.

Macroscopically, then, the upper boundary of the conus terminalis can be determined only arbitrarily, or not at all. But it can be accurately determined by histological examination. In the third sacral segment, the character of the transverse section is considerably altered. The gray substance, in comparison with the white, is excessively developed. As Fig. 91 shows, the plump, broad anterior horns are abruptly rounded

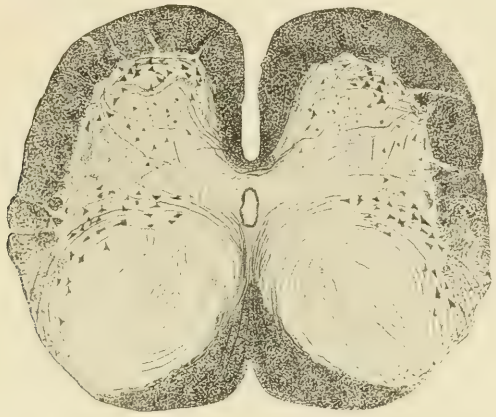


FIG. 91.—Cross-section through the conus terminalis.

anteriorly, and no longer have any lateral protuberance, as they still have in the upper sacral cord; the posterior horns fairly bulge, due to the luxuriant development of the gelatinous substance. The ganglion cells in the anterior horns are very scanty here, but they are all the more numerous in the transitional zone between anterior and posterior horns; from here, fibres radiate directly into the postero-lateral columns, and into the roots that are there situated. The lower the point in the conus from which the sections are taken, the larger and more powerful are the ganglion cells in the "intermediary zone." In the arrangement of the white substance also, essential differences may be established in the conus from the other parts of the spinal cord. At the same level of the cord at which the great ganglion cells disappear from the anterior horn and new groups of them appear in the intermediary zone, one finds that a bundle of medullated fibres is seen to pass directly from the posterior columns towards the front, that is, ventrally towards the central canal and on both sides, increases the fibres in the intermediary zone. Here, then, and this characterizes the beginning of the conus, no posterior gray commissure of the gray substance can be found.

Transverse section above the conus shows the degeneration in the lateral pyramidal columns does not extend downwards beyond the third sacral segment. On the other hand, in the posterior columns of the sacral cord and in its lowest part, in the conus alongside the septum medianum posticum course very many *descending* conducting paths, whereas in the cervical, dorsal, and lumbar segments, these tracts contain almost only centripetal and ascendingly degenerating fibres, and it seems to be these descending fibres, which then radiate towards the front into the gray substance, and are lost in the intermediary zone.

In the conus medullaris the centers for the sexual functions and for the evacuation of bladder and rectum were always supposed to have been situated, but more recent investigations cast a great deal of doubt upon the correctness of this hypothesis. Thus, it can be demonstrated, that the act of parturition may proceed normally, even after disjunction of the lowest section of the spinal cord. Likewise it is certain that after the lower sacral cord is completely destroyed, erection and ejaculation of the semen into the urethra may still take place; as a result of the paralysis of the ischio- and bulbo-cavernosi, however, the semen can no longer be ejaculated from the urethra, but dribbles slowly away.

Neither can it be doubted any longer that the last nervous centers subserving the bladder and the rectum are situated outside of the spinal cord in the sympathetic nervous system; it must be remembered that this theory concerns organs with involuntary musculature and such are not innervated directly from the cerebro-spinal system anywhere in the body. It is true that the reflex, which subserves evacuation of urine and feces, can be carried out voluntarily to a certain degree. But the formerly generally accepted hypothesis, that one center for the detrusor and another for the sphincter vesicæ are situated in the conus medullaris, is probably incorrect. On the contrary, we must conceive of the automatic process, which leads to relaxation of the sphincter and to contraction of the walls of the bladder, as being entirely excited in the sympathetic system. We cannot decide, as yet, by what paths this reflex is evoked, whether by way of the rami communicantes over the ganglion mesenter inferius and ganglion hypogastricum, or whether possibly this process might be aroused by a contraction of the striped musculature situated in the pelvis.

The conditions underlying the evacuation of the bowel-contents are much clearer. By abdominal pressure, the column of feces is pushed downward somewhat; then the peristaltic movement of the ampulla recti takes hold and this leads to the ejection of the excrement. It is very improbable, that in this process, a center for defecation in the conus is actively concerned. True it is that the transversely striated sphincter ani externus has its ganglion cells in the lower division of the spinal cord, just as has the compressor urethræ, which is also under the influence of the will-

power; spontaneous peristaltic movements of the rectum and the musculature of the bladder may therefore be repressed to a certain degree, i. e., it is possible to retain urine and feces, even when desire to evacuate exists. In like manner the centripetal paths, over which we are informed as to the degree of fulness in the bladder and the ampulla recti, as well as about the processes of motility taking place there, probably pass through the conus terminalis. Thus is explained, that when the lower sacral cord is diseased or the nerve bundles in the cauda equina passing thither are interrupted, the patient is not aware of the fulness of the bladder and rectum, and is not able to perform the automatic process necessary for the ejection of the excrement (retentio urinæ et fæcium). Nor is he able to inhibit the reflex, once it has started, as in diarrhœa or in spontaneous evacuation of urine by contraction of the voluntary musculature of the sphincters of the anus and the bladder (incontinentia urinæ et alvi). The disturbances in micturition and defecation in affections of the conus and the cauda equina do not differ from those which develop in connection with transverse affections in the rest of the spinal cord. First there is always retention of urine and feces, later there appear involuntary automatic evacuations.

The diseases of the conus medullaris are often accompanied by those of the upper sacral and the lumbar portion of the cord, or they attack the bundles of roots which spring from these parts and surround the conus; it is therefore advisable to give a short summary of the situation of the *nuclear areas* of the individual muscle groups in the lumbar enlargement of the spinal cord. But we must remark, that here only the *center* of a nuclear realm is indicated, the nuclear realms of the larger muscle groups being mostly extensive and reaching over several segments.

1. Lumbar segment: lower part of the abdominal muscles, quadratus lumborum.
2. Lumbar segment: iliopsoas, cremaster, sartorius.
3. Lumbar segment: adductors of the thigh.
4. Lumbar segment: quadriceps femoris, tibialis anticus.
5. Lumbar segment: abductors of the thigh (glutæus medius and minimus), flexors of the knee (semimembranosus and semitendonosus and biceps).

1. Sacral segment: glutæus maximus, outward rotator of the hip, extensors of the toes.
2. Sacral segment: gastrocnemius, soleus, peronei, flexors of the toes.
3. Sacral segment: ischio- and bulbocavernosus, perineal muscles (compressor urethræ).
4. and 5. Sacral segment: sphincter ani externus, levator ani.

Figures 92–98[1] show what *disturbances in sensibility* correspond to

[1] Figs. 92–98 are taken from a previous work of the author's (*L. R. Müller*, Investigations into the anatomy and pathology of the lowest segment of the spinal cord. Deutsche Zeitschrift fuer Nervenheilkunde, vol. xiv).

the diseases in the various segments of the lumbar enlargement. As one may see from Fig. 98, the nerves from a small oval zone about the anus reach the fourth sacral segment. In a lesion of the third sacral segment, the anæsthesia extends also, to the perineum, the dorsum of the penis and to the middle parts of the scrotum; in a lesion of the second sacral segment the whole external genital organs are anæsthetic, the insensitive zone in the buttocks has become considerably larger, extending to the upper portion of the posterior aspect of the thigh; there is also a large insensitive area on the outer portion of the foot. In lesions of the first sacral segment, this spreads to the outer and posterior portion of the leg proper, which is supplied by the peroneus nerve. Transverse diseases of the fifth lumbar segment lead to

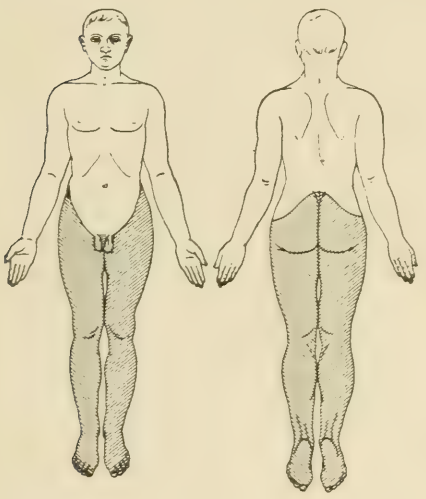


FIG. 92.—Lesion at the level of the 2d lumbar segment.

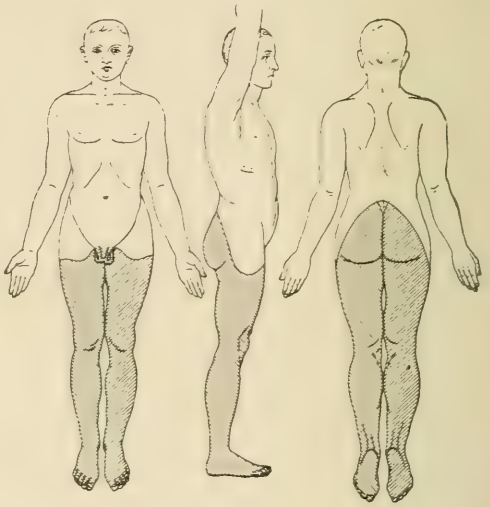


FIG. 93.—Lesion at the level of the 3d lumbar segment.

the typical "riding breeches" anæsthesia (Reithosenanæsthesie). In all these disturbances of sensibility, the testicles in the anæsthetic scrotum, since they are innervated by the spermatic nerve from the second lumbar segment, are still very sensitive to pressure.

Stroking the skin in the immediate vicinity of the anus causes contraction of the sphincter ani and the levator ani. This reflex, the *anal reflex*, has its center in the conus terminalis, hence may still be elicited in diseases of the upper sacral cord. But the Achilles tendon reflex is lost in a disturbance at this level since it has its seat in the region of the epiconus, in the second sacral segment. The patellar reflex must be localized much higher still, in the fourth lumbar segment.

As has been mentioned before, the lower end of the conus terminalis reaches only to the upper margin, or at most, to the middle of the second lumbar vertebra; from that point on the fibre bundle of the cauda equina,

which is about as thick as a finger, contains only the filum terminale. The parallel strands of the cauda equina must pass for a long way inside the spinal canal, in order to reach their foramina of exit. Thus the first sacral nerve arises at the level of the first lumbar vertebra and leaves the dural sac

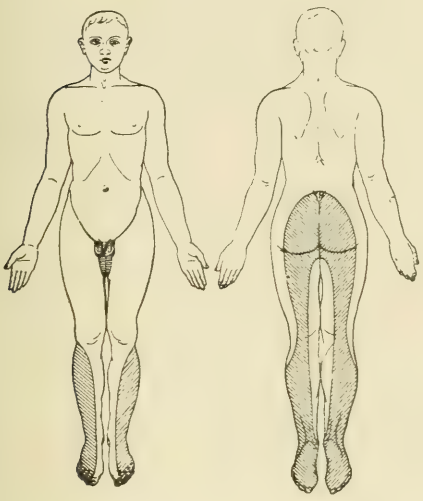


FIG. 94.—Lesion at the level of the 5th lumbar segment.

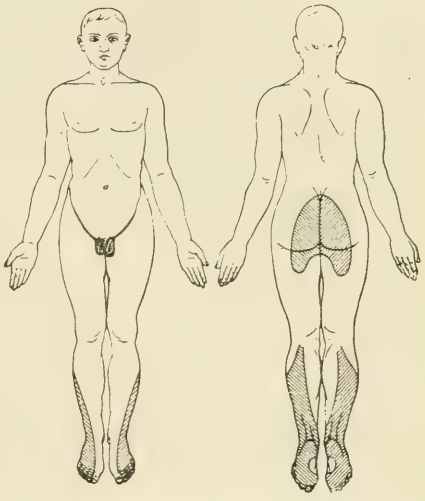


FIG. 95.—Lesion in the 1st sacral segment.

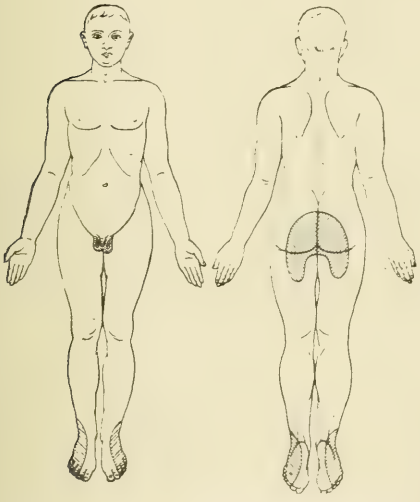


FIG. 96.—Lesion in the 2d sacral segment.

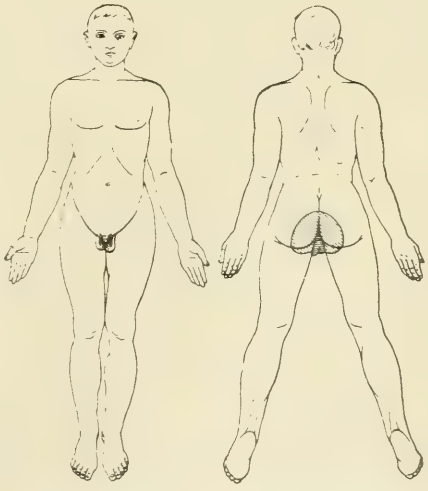


FIG. 97.—Lesion in the 3d sacral segment.

only in the sacrum, in order to enter the pelvis from the first sacral foramen. Therefore, it must course a distance of 14 cm. within the dural sac of an adult. A cross-section through the cauda equina, at the level of the conus terminalis strikes besides all the sacral nerves, all the lumbar roots also with the exception of the first. Naturally, the lumbar roots which escape

higher up, are situated at the outer side in the bundle of the cauda equina, while the sacral fibres which escape farther down, are medially situated.

The sensory roots form their spinal ganglion only after their escape from the dural sac; it reaches a quite considerable size in the lumbar nerves, but especially so in the sacral nerves. Now, finally, after the sensory root has passed through the spinal ganglion, it unites with the anterior roots to form the *peripheral nerve.* The fibres of the cauda equina, therefore, are not, as often happens, to be regarded as peripheral nerves, but as root fibres, i. e., in the cauda, the motor and sensory bundles course *separated* from each other; we may easily convince ourselves in processes in which only the sensory tracts degenerate (for instance in tabes) that these sensory bundles are situated all together, in one posterior field, while the motor roots are

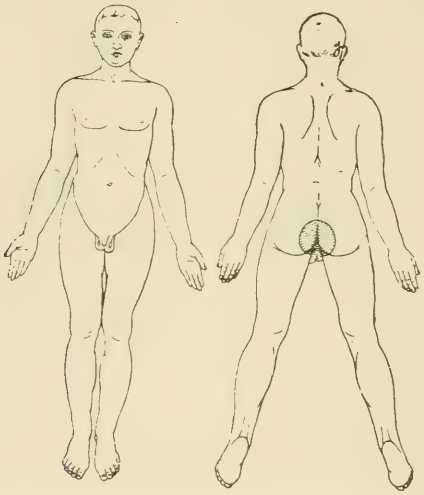


FIG. 98.—Lesion in the 4th sacral segment.

situated farther front, also joined in groups. It is clear that this fact is of great importance for the diagnosis and localization of the disturbances with which we are here concerned (caudal diseases).

After the escape of the caudal fibres from the spinal canal one distinguishes *five pairs of lumbar nerves*, the first of which pass out between the first and second lumbar vertebræ, the last between the fifth lumbar vertebra and the sacrum; *five pairs of sacral nerves* which radiate outwards through the foramina sacralia anteriora and posteriora and finally one coccygeal nerve, which after passing the hiatus sacralis courses downwards. Now, while the intercostal nerves form no plexus, the outgoing lumbar and sacral nerves are *mingled,* each in a plexus, and now for the first time they separate into the real peripheral nerves. Thus, the distribution of fibres in the cauda equina and the order of their origin from the spinal cord is different from the arrangement in the peripheral nervous system. The *plexus lumbalis* is formed by the four

upper lumbar nerves, the *sacral plexus*, the large plexus situated on the anterior surface of the pyriformis muscle, is made up from parts of the fourth, from the fifth lumbar nerve and from the upper four sacral nerves. The last sacral nerve and the *coccygeal nerve* unite to form the plexus coccygeus. The nervus ano-coccygeus thus formed supplies the skin in the neighborhood of the tip of the coccyx.

To summarize from the innervation of the pelvis, what is important for diagnosis, we must point out that the anterior and external portions of the pelvis and thigh get all their sensory supply from the lumbar plexus; only the penis and the scrotum are supplied by the pudendal nerve, which springs from the sacral plexus. The posterior portion of the pelvis, also, or, better, of the buttocks, is innervated by the ramus iliacus of the nervus ilio-hypogastricus and by the nervus cutaneus femoris lateralis from the lumbar plexus; on the other hand, an oval zone, about the size of a saucer, lying around the anus, and the perineum gets its sensory supply from the sacral plexus and the coccygeal plexus.

The conditions *within the pelvis* are still more complicated. Of the muscles there situated, the psoas, the iliacus internus, the obturator externus, the *cremaster* and the tunica dartos are innervated from the lumbar plexus, all the other small pelvic muscles (M. pyriformis, M. obturator internus, Mm. gemelli), the musculature of the perineum, the anus, and the large muscles of the buttocks receive their nerves from the sacral plexus.

The *diseases of the conus terminalis* are brought about by various causes. Since the conus terminalis is part of the substance of the spinal cord, all pathologic-anatomical processes, which attack the rest of the medulla spinalis, may be localized occasionally in this, its lowest part, as well. As a matter of fact, isolated cases of myelitis, hæmatomyelia, multiple sclerosis, and various sorts of tumors have been described in the conus medullaris. In the great majority of cases, however, *traumatisms* are responsible for the affections of the conus. The first lumbar vertebra, behind which the conus is situated—as a result of the static relations of the spinal column, in its capacity as a transitional vertebra from the rigid, backwardly curved dorsal spinal column to the movable lordotic lumbar spinal column—suffers with especial frequency compression fracture from violent jolts, i. e., falls upon the legs or the buttocks. By such an injury, the spinal canal is narrowed and the lowest section of the spinal cord crushed. The fibres of the cauda equina, which surround the conus, being far tougher and more capable of resistance, usually do not suffer, when the first lumbar vertebra is broken, since they are able to get out of the way in time, so that pure cord lesions without participation of the roots have been observed. Fig. 99 shows the cross-section of a dural sac, broadly flattened out by pressure from a compression fracture of the first lumbar vertebra; the cauda bundles are very well preserved, the destroyed conus is replaced by gliar and fibrous scar tissue. The preparation is taken

from a patient, who by falling from a height of 10 m. sustained a fracture of his first lumbar vertebra.

Occasionally, after a concussion, the typical symptoms of a conus affection appear, though it is impossible to discover the presence of a deformity in the spinal column. A possible explanation for such cases might lie in the fact that there had been no fracture, but merely a momentary elastic displacement of the upper lumbar spinal column, which, however, sufficed to crush the conus. On the other hand, the following hypothesis has been advanced by *Fischler:* The fall is followed by excessive forward flexion of the trunk, which in turn, causes violent bending in the spinal cord, so that, at the place where the caudal roots enter the conus, there may be lacerations and secondary hemorrhages into the cord.

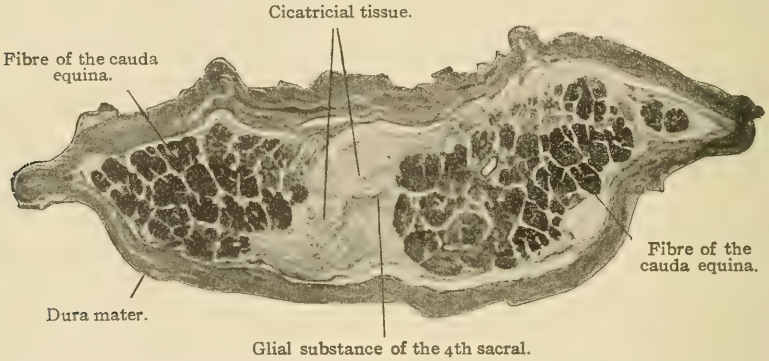


FIG. 99.—Cross-section through the dural sac after crushing of the 1st lumbar vertebra.

Besides traumatism, the other causes of disease play but a slight part. Now and again the symptoms of an affection of the conus develop spontaneously, and with comparative rapidity, so that the existence of a myelitis must be suspected. Only rarely do tumors develop in the conus itself; on the other hand, it occurs more frequently that the conus is compressed by tumors starting in the cauda equina or the vertebræ, by pachymeningitis, or by tuberculous processes. The literature does not as yet furnish cases of syringomyelia, poliomyelitis, sclerosis multiplex of the conus medullaris, which were positively identified as such by postmortem examinations.

The **clinical phenomena** of conus diseases are limited to an anæsthesia over the sacrum, anus, perineum, in the genitalia, and to a disturbance in the functions of the rectum, the bladder and the sexual organs; nearly always there appears, as has been already stated, retention of urine and feces. After ischuria paradoxa has persisted for some weeks, or has been combated by catheterization, frequent spontaneous evacuations of urine occur. At a certain degree of fulness in the bladder, automatic evacuation is induced, Shortly before, the patients usually have a certain indefinite sensation in the

abdominal region, so that, if a receptacle is at hand, they may avoid getting soiled by the urine. But voluntary excitation or suppression of micturition is and remains impossible. Closure of the sphincter is, however, not so secure as in the healthy, so that during coughing, sneezing, or more violent movements, urine generally escapes. Retention of feces, on the other hand, remains permanently; usually, there is evacuation only after the administration of a purgative. After taking the purgatives, the patients must remain for hours on the toilet, because they have no sensation of the oncoming defecation, and they can not hold it back even for a moment because of the paralysis of the external sphincter ani. Since the patients are not able to retain rectal injections, the constipation can not be combated by enemata. The paralysis of the sphincter ani externus is also evinced by the fact that the anal reflex is always lost in affections of the conus. The sexual functions, immediately after an injury to the conus, and as long as severe general phenomena like ischuria or acute cystitis persist, are entirely absent. Later on, in spite of the disjunction of the lower section of the spinal cord, penile erection may occur and evacuation of the semen into the urethra, though, as a rule, the sensory impressions and orgasm are absent. From the urethra, the semen dribbles but slowly; hence it is always possible that such patients may have fructifying cohabitation.

In *pure* affections of the conus, i. e., if the pathologic-anatomical process is restricted to the lowest three sacral and the coccygeal segment, there are no disturbances in sensation or phenomena of paralysis in the lower extremities. Locomotion therefore is in no way interfered with. Very frequently, however, the upper sacral cord, or even the lowest lumbar cord, are simultaneously affected, and then there are added to the bladder and rectal disturbances, paralyses of the peronei and flattening and functional loss of the glutæus maximus and of the flexors of the knee; the loss of sensation about the anus, of the perineum and the genitalia, are supplemented by the anæsthetic zones, represented in Figs. 92–98.

After traumatic lesions of the conus, the morbid changes in the cord, such as hemorrhage, serous infiltration, edema, or reactive inflammation extend nearly always to the middle of the lumbar enlargement. In time, however, the reparable disturbances, and together with them, the phenomena of paralysis and the loss of sensation disappear in the lower extremities, and only those symptoms remain which were excited by crushing the substance of the cord.

Minor denominates as the *epiconus* that part of the lumbar enlargement which is situated immediately above the conus and corresponds to the 5th lumbar and the 1st and 2nd sacral segments. Disease pictures are described by this author, in which the paralysis is confined to the realms of the peronei and glutæi, so that even the knee reflexes, on the one hand, and the control over the sphincters of the bladder and rectum, on the other, are

retained. Such a symptom-complex is nearly always caused by acute poliomyelitis.

In transverse diseases in the *lumbar cord*, caused, as they may be, by inflammatory or traumatic myelitis, or by tumorous formations, there results, from the interruption in conduction, also paralysis of the muscular groups innervated by the sacral plexus. But here the type of paralysis will be spastic (ankle clonus, retention of the Achilles reflex) whereas in the realm of the lumbar plexus, degenerative, i. e., atrophic paralyses may be demonstrated (absence of the patellar tendon reflex).

If the motor disturbances predominate in the diseases of the conus, the sensory irritation phenomena are characteristic of the *caudal diseases*. Scarcely any other disease is associated with such painful conditions, as the gradual compression of the bundle of the cauda equina, as it results from tumors. The pains are referred to the anus, the perineum, and the genitalia, and frequently radiate to the legs proper and the peroneal realm. In advanced cases, the corresponding cutaneous areas are insensitive, as a result of the break in conduction in the roots of the cauda, but nevertheless there are still pains in this region, and the condition is then spoken of as "anæsthesia dolorosa."

The disturbances caused by caudal diseases appear gradually, especially when they are caused by tumors. At first, there are pains only with certain movements of the body, or by pressure on a vertebra; later they appear spontaneously, become almost continuous, and are increased to unbearable violence.

Although the cauda equina contains, in its upper part, roots which come from the lumbar cord, nevertheless, the pains in compression of the cauda equina are nearly always restricted to the realm of the sacral roots. This fact, taught us by experience, is not easily explained, since it is only natural to suppose, that in a space-contracting process in the canal of the lumbar spinal column the roots, situated at the outer side would be affected likewise.

In the cauda equina, as was stated above, the sensory and motor tracts are still separated. Thus it is comprehensible, that sensory irritation phenomena may exist without motor disturbances. Pareses and paralyses develop much later, if at all. The paralyses are flaccid and usually not strictly symmetrical, i. e., more pronounced on one side than on the other. The disturbances in bladder and rectum caused by caudal affections are in no wise different from those which appear in connection with conus diseases, except, possibly, that their onset is more gradual; before ischuria supervenes the evacuation of the urine is made very difficult for a long time.

As cause of their origin, traumatism in lesions of the cauda equina plays not nearly so important a part as in those of the conus. In the first place, the lower lumbar vertebræ and the os sacrum, are not so frequently exposed to

fractures as the first lumbar vertebra, which is especially endangered in falls on the legs, or on the buttocks; then the tougher nerve roots, which are more capable of resistance, evade injury in compression of the spinal cord more easily than can the tender substance of the sacral cord. Very often, however, *tumors* cause injuries of the caudal roots. All sorts of growths, like neuroma, lymphangioma, fibromyxoma, enchondroma, exostoses, cysticerci, etc., may develop within the lower spinal canal, inside and outside of the dural sac. But especially frequently metastases lead to compression of the caudal fibres and above all carcinomata and sarcomata show an inclination to secondary tumor formation in the lumbar spinal column, and in the os sacrum. On the other hand, tubercular and gummatous processes occur far more rarely in the lower part of the vertebral column than in the dorsal or cervical parts.

It is to be remembered that tumors *outside of the spinal canal* may also lead to injury of the nerve tracts here concerned. A tumor located in the pelvis, and attached to the sacrum will, as it grows, attack the fibres which pass to the bladder, the genitalia, and the rectum. Such disturbances assume the character of *plexus paralyses*. For the nerve tracts are not usually affected by the injury, at a place where the bundles are already leaving the plexus to separate into single nerves, but are injured immediately after they leave the spinal column, that is, in the plexus, or before they have joined to form it.

Finally it must be mentioned that diseases of the *peripheral nerves* present similar clinical phenomena as affections of the plexus or cauda. Frequently, cases of sciatica, or of coccygodynia can scarcely be distinguished from the initial difficulties caused by compression of the fibres of the cauda equina, or of the sacral and coccygeal plexuses.

From the facts here presented, one may appreciate what difficulties **differential diagnosis** may present, in cases in which one must decide whether the lower portion of the spinal cord itself, or its surrounding and outgoing roots, in their long course, or, finally the plexus and its peripheral processes are injured. But differential diagnosis has not only theoretical interest, but is of great practical importance, also. Thus, the prognosis changes considerably, as the location of the seat of the disease varies. In a sciatica, the prognosis is usually favorable; if the same subjective disturbances are caused by the formation of a tumor in the lower spinal canal, one must count upon an aggravation of the disease. Diseases of the lower spinal cord itself usually run their course rapidly, but their resulting phenomena remain stationary, after the reactive phenomena have disappeared, i. e., improvement and cure are excluded, but neither is an aggravation of the disease to be feared, unless complications appear through decubitus, or through cystitis and pyelitis. In our discussion of differential diagnosis, we must emphasize again the fact, that in diseases of the cord itself, the motor phenomena of functional loss predominate, that these are usually symmetrical and exhibit a

segmentary arrangement. The sensory disturbances, in diseases of the conus, are restricted to *loss* of sensation; the *anæsthesia* is frequently *dissociated*, i. e., includes only certain qualities of sensation, such as pain and temperature. The *Brown-Séquard* symptom complex points definitely to a localization of the disease in the lumbar enlargement. Furthermore, the sensory *irritation* symptoms are lacking in conus diseases. These, however, predominate in the clinical picture, when a caudal affection is concerned. If the pains are permanently unilateral, one must consider whether a plexus disease, or a neuralgia of a peripheral nerve, f. i., ischialgia, is not present. Sensitiveness of the musculature to pressure speaks for neuritis; in the latter case, the paralyses are usually definitely located in the area of one or more peripheral nerves. The disturbances in the bladder and rectum can not be used in a differential diagnosis between "conus" and "cauda." Only this one fact points to a caudal affection, if together with ischuria, and retentio fæcium, violent pains radiate to the perineum. In diseases of the peripheral nerves, as in coccygodynia, in sciatica, or polyneuritis, disturbances in evacuation of urine and feces are always lacking.

In typical cases, the differential diagnosis between diseases of the cauda and conus presents no difficulties too great to be overcome. But if a caudal lesion had been diagnosed, then the difficult decision must be made, at *what height of the cauda* the lesion is to be sought. The rule may be established here, as in diseases of the spinal cord, to look for the disturbance at as high a point as possible. The nerves, which subserve the bladder and rectum, may be injured throughout their entire course through the cauda, and it has been shown several times, that in diseases of the cauda located high, these nerves situated in the middle and passing out farthest down, usually suffer *first*. . The lesion of the cauda must at least be localized at a height, which corresponds to the exit of the uppermost roots, in which irritation phenomena are still demonstrable.

If a disease of the cauda equina and the conus medullaris are *synchronously* present, a more exact diagnosis will usually be impossible.

Treatment is absolutely impotent when facing myelitic processes in the lower division of the spinal cord, whether they be of inflammatory or traumatic character, whether they be recent or partly healed. Likewise, when a tumor has caused pressure atrophy of the cord, no result is attained by its removal.

Conditions in the cauda equina are different. There, by extirpation of the compressing tumor, an amelioration of troubles, indeed, under certain conditions, complete recovery may be effected. The motor roots of the cauda have their trophic centre in the spinal cord, and the sensory, in the spinal ganglia, and thence, a restoration of fibres, and at the same time, of the functions may result. The operative technique also is not exceptionally difficult. But permanent results are opposed by the fact discussed

above, that in the lower spinal canal, we rarely have to deal with isolated tumors; on the contrary, it is usually a matter of metastases. Operation is absolutely hopeless, if tumor masses penetrate the vertebral bones. Naturally, with the first symptoms of a caudal compression, treatment with iodide of potassium and mercury will always be instituted at once, assuming the possibility of a gummatous change or a pachymeningitis specifica of the lower dural sac being present.

IV

THE MYOPATHIES

WITHOUT DEMONSTRABLE CHANGE IN THE NERVOUS SYSTEM

BY

HANS CURSCHMANN (Mainz)

Under the term *myopathies*, we can group together a series of disease pictures, all of which are of rather rare occurrence: dystrophia musculorum progressiva (Erb), myotonia congenita (Thomsen), myasthenia pseudo-paralytica and lastly a disease, that has been known only for a few years—congenital myatonia (Oppenheim). All four are severe afflictions of the function of movement; some forms are accompanied by trophic disturbances of the musculature (dystrophy, amyotrophic myotonia), others run their course without any macroscopic changes in it (myasthenia, myotonia congenita, myatonia (Oppenheim). Their disturbances in movement vary considerably; in part they are directly antagonistic (myotonia and myasthenia). In one respect, however, all four diseases are alike: in the macroscopic and microscopic *intactness of the nervous system*, on the one hand, and in the well-characterized macroscopic, or, as is mostly the case, only microscopic *changes in the musculature*. It has been attempted, also, to include the first three diseases under one pathogenetic hypothesis (Lundborg) without, however, being able to prove their common parathyreogenic origin.

1. DYSTROPHIA MUSCULORUM PROGRESSIVA (ERB)

From the confusion of the various forms of muscular wasting that still reigned in the time of *Friedreich*, *Duchenne*, *Charcot* and others, *Erb's* searching hand has extricated, under the above name, a definite group of *pure myopathic* atrophies and pseudo-hypertrophies. Even though the usual division into various sub-forms (infantile, pseudo-hypertrophic form, juvenile form, etc.), is to be retained in these diseases, the fact must nevertheless be emphasized, that transitional forms occur quite as frequently, as the pure and separate forms, and this always brings the *unity* of *Erb's* disease picture to clear recognition. One may therefore well dispense with distinctions between still more separate forms (*Leyden-Moebius*, *Duchenne-Déjérine*, etc.).

Definition.—*Erb's* dystrophy embraces forms with the following peculiarities: 1. *Beginning of the disease in childhood* and youth (from earliest

infancy to the end of the second decade, very rarely in the thirties and forties); 2. very frequent (but by no means constant) *familial appearance* in such a manner that the disease is usually found in *several brothers and sisters*, but there is but rarely transmission from ancestors to descendants; 3. atrophies and pseudo-hypertrophies regularly attack definite groups of muscles, the atrophies usually definite muscles of the chest, shoulder-girdle, back, arms, buttocks and thighs, often, too, those of the face, while *the distal parts of the limbs are nearly always spared* to the elbow and to the knee (in striking contrast to spinal muscular atrophy); the pseudo-hypertrophy prefers definite muscles of the arm and above all, the muscles of the calf of the leg; 4. *the clinical signs of degenerative atrophy* (fibrillary tremors, electrical and mechanical reactions of degeneration) are nearly always *absent;* 5. the following other *symptoms of functional loss of the central and peripheral neuron* are always *absent, also:* participation of the sensory cerebral nerves, reflex augmentation (*Babinski*) with spastic contractures, changes in sensibility and in the activities of the sphincters; 6. *anatomical changes are found regularly in the muscular apparatus*, but in pure forms of the disease *never in the nervous system.*

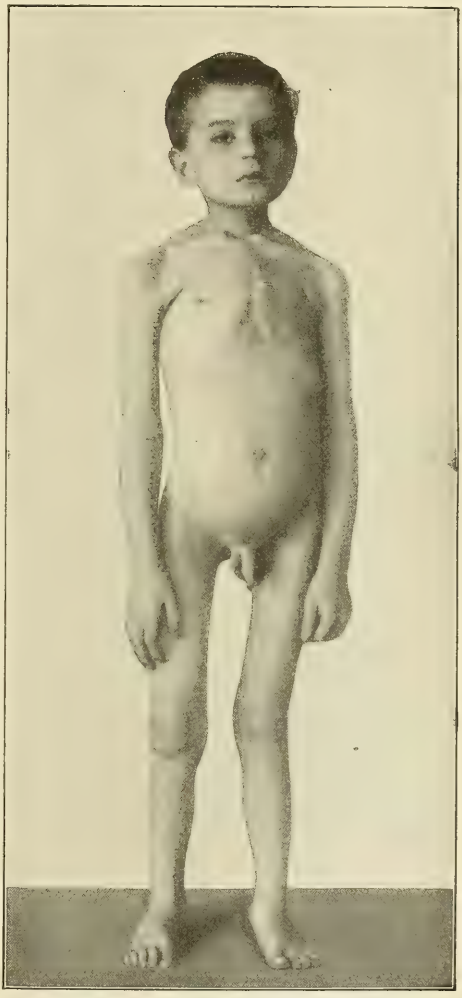


FIG. 100.—Infantile atrophic form of dystrophy with participation of the face. (*After Heinr. Curschmann.*)

The **infantile atrophic form of dystrophy** is relatively the most frequent; its course may run with and more rarely without involvement of the face; pseudo-hypertrophies are scarcely ever absent. The disease begins (it is more frequent in boys than in girls) insidiously and very slowly in early childhood between the third and fifth years, after the patients have already learned to walk in the normal way. The first things the parents notice are: a somewhat slower, waddling and rocking gait, difficulty in running, jumping and especially in rising from a recumbent or bent-over position. Simul-

taneously, a lordosis of the spinal column develops. As a rule, disturbances in the motility of the upper extremities (in raising the arms and shoulders) are not perceived till later. At this time paralytic phenomena in certain facial muscles also appear quite frequently. After having existed several years the disease reaches its climax.

The findings are now usually as follows (Fig. 100): The most serious disturbances nearly always affect the *musculature* of the *trunk, pelvis and thigh.* The gait is noticeably uncertain, rocking, waddling (because of the weakness of the glutæi medii, *Strümpell*), the thighs are raised laboriously and relatively high, the feet fall stamping to the ground (weakness of the muscles of the thigh). In walking and standing a pronounced lordosis of the lumbar spinal column appears, which may attain grotesque degrees, a result of the atrophy of the muscles of the back and pelvis; the abdomen protrudes considerably. The atrophies constantly single out, besides the long muscles of the back, the glutæi, the extensors of the leg proper (quadriceps), somewhat less frequently, its flexors. The paralysis of the two former causes the well-known phenomenon in rising from a recumbent position (cf. Fig. 102): first the children by means of their arms raise themselves high enough to stand on "all fours," then they climb up on themselves, as it were, resting their arms first upon their knees and then upon their thighs.

Associated with the atrophies, *pseudo-hypertrophies* are nearly always found in the lower extremities, most constantly in the musculis gastrocnemius; less frequently traces of pseudo-hypertrophy are found in the glutæi and the deltoid. The hypertrophic muscles usually feel more flaccid than normal muscles, often peculiarly doughy, sometimes, however, dense and solid. Their strength is often normal, somewhat less frequently diminished, very rarely increased.

In the infantile form the muscles of the shoulder-girdle and the arms are for a long time less severely affected than those of the trunk and the legs. But atrophies of the m. pectoralis, the m. cucullares, the m. serrati and others are quite constant. The resulting disturbances in motility, which especially affect the raising and use of the arms, when they are lifted above a horizontal position, are discussed in greater detail under the juvenile form of the disease, in which they play a far greater part. The forearm is nearly always spared in the infantile form.

The face is attacked by the atrophy and paresis at an early period. First is noticed the weakness of the orbicular muscles of the eyes and the mouth; the closure of the eyes is performed incompletely and lacks power; the mouth can not be pursed. As a result of atrophy of its muscles, the upper lip protrudes abnormally; there develops a suggestion of "tapir-snout." Later the atrophy extends—not always symmetrically—over the musculature of the cheeks and forehead, so that finally the myopathic mask-face appears. The tongue and pharynx, as well as the muscles of mastication—in contrast

to bulbar paralyses of other genesis—nearly always remain intact; likewise the external and internal muscles of the eye.

It has already been stated that the sensory cerebral nerves, sensibility and sphincters remain normal. Likewise, psychic anomalies belong to the exceptions; on the contrary, the children, as a rule, are intellectually alert and clever.

A rarer modification of infantile dystrophy is **pseudo-hypertrophy of the muscles,** described first by *Griesinger,* later by *Duchenne* and others. It also begins in earliest childhood, between the third and sixth years; its

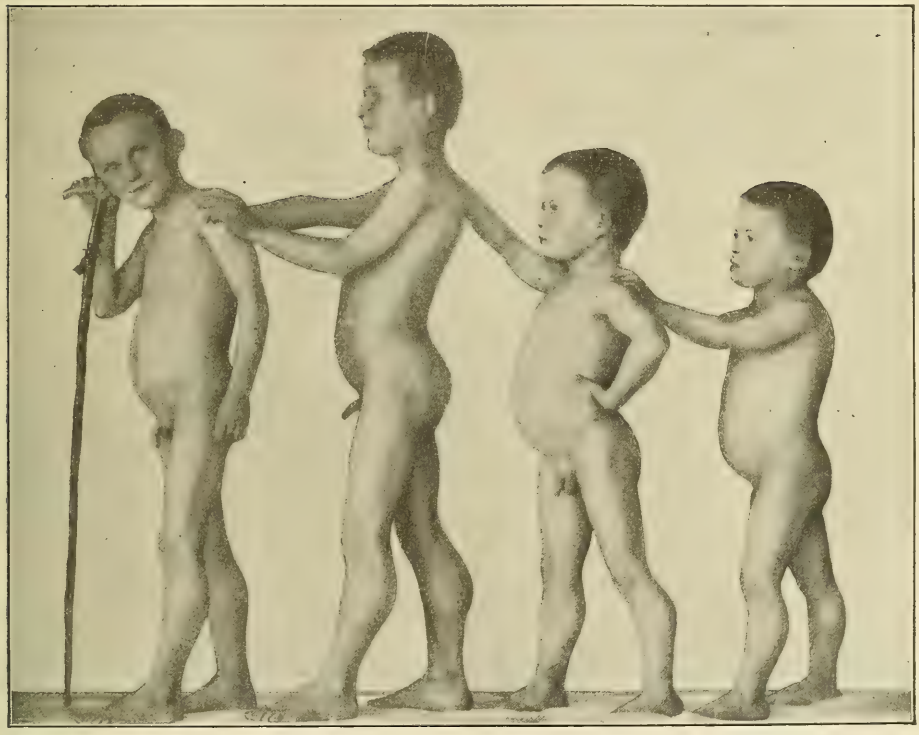


FIG. 101.—Four children of the same parents with infantile muscular dystrophy. The two older children show the atrophic, the two younger boys, the almost pure hypertrophic form. (*After Heinr. Curschmann.*)

appearance is very insidious, since the general stoutness of the body does not, at first, impress the parents, as the sign of a disease.

In typical cases of this disease the pseudo-hypertrophies overshadow the atrophies decidedly. The muscles of the calves and thighs, often also of the buttocks, stand out with enormous volume; in like manner, the muscles of the shoulder and arm (especially the m. triceps) become hypertrophic. At the same time the abdomen is protruded, because of the existing lordosis, and seems abnormally developed. The external picture might resemble that of a caricature of a youthful Hercules.

At the same time, the consistency of the hypertrophied muscles is often very soft and spongy, but occasionally, however, as in genuine hypertrophies, solid. Correspondingly, the functioning power of these muscles is, for the most part, relatively reduced; but we have also seen cases in which hypertrophied muscles were abnormally strong. This is true especially of the musculature of the calf, the endurance and strength of which made one of my little patients, a virtuoso in "toe-running."

Beside the pseudo-hypertrophies, atrophies also are nearly always discovered upon closer examination; they attack preferentially the long muscles of the back, and not infrequently also the glutæi. The weakness of the pseudo-hypertrophic and atrophic muscles of the trunk and the legs now cause disturbances in posture (especially in standing and walking) and in the gait itself, which closely resemble those of the atrophic dystrophies; pronounced lumbar lordosis and waddling, rocking outward walking. The ability of the patient to raise himself from the ground is always disturbed. The weakness of the arm and shoulder muscles appears less distinctly in the pure pseudo-hypertrophic forms, though it is never altogether absent. Atrophy and paresis of the musculature of the face are said not to occur in these forms.

Concerning the occurrence and course of pseudo-hypertrophy the fact must be emphasized, that not infrequently it is found in families, in which the older children suffer from atrophic dystrophy, as the reproduced dystrophic brothers from the Leipsic clinic show in a classical way (Fig. 101). In such cases the pseudo-hypertrophy usually passes over into the atrophic form with involvement of the face. In other instances, also, transitions between the two forms frequently occur.

The juvenile form of dystrophy, which is almost as common as the infantile atrophic form, does not show familial appearance so frequently as the forms just described. Sporadic cases are certainly no rarities. The disease attacks, as *Erb*, *Strümpell* and I have observed, the female sex relatively more frequently than do the infantile forms.

The onset in the juvenile form occurs about the middle of the second decade and is usually most insidious, even though more rapid progress has supposedly been observed after *traumatism.*

As characteristic distinctive signs from the forms discussed heretofore, we must mention first and foremost: *the prevalence and the early beginning of the disturbances in the shoulder-girdle and the upper extremities and the falling into the background of the pseudo-hypertrophy in the juvenile form.* It is not improbable that the former characteristic, lacking in the infantile form, is to be explained by the considerably increased demands that are being made upon the muscles of the arms and shoulders (in the sense of the "using up" theory of *L. Edinger*).

The entire type of the patients differs from that of the infantile form by

FIG. 102.—*a–e.* Rising from the ground in dystrophia musculorum. (*After Heinr. Curschmann.*)

FIG. 102, *b.*

FIG. 102, *c.*

FIG. 102, *d.*

FIG. 102, *e.*

the pronounced changes in the form of the shoulders, thorax and arms (Fig. 103). Owing to the paralyses of the m. cucullares, of the muscles that fix the scapula and in part of the m. pectorales, the shoulders sink far downwards and frequently not only downwards but forwards as well. From these "fallen" shoulders, arms hang often thin as broom-sticks, and at their top lies a deltoideus of almost normal volume which has also slipped downwards and forearms, which, by atrophy of the supinator longus and occasionally of other muscles, have lost their oval contour and acquired an approximately round one. Through the overhanging of the shoulders and the atrophy of the pectoral muscles, which causes the breast-arm furrow to run upwards instead of downwards, there appears a sunken breast form, the "thorax en bateau," occasionally an actual funnel-shaped thorax. Especially striking are the changes in form and disturbances in function, which are produced through the defects of the scapular muscles. Through paralysis of the serratus, the scapulæ protrude like wings, not only while at rest, but especially when lifting; in addition, through the disappearance of the m. cucullares, the shoulder-blades are in an abnormally low position and far from the middle line. If the attempt is made to raise the patient by or under the upper arms, this fails, as the shoulders are "loose," cannot be fixed, and during the attempt are pulled limply upwards above the ears. If the patient is ordered to press the arm, that has been raised to a horizontal position, downwards against a resistance, there also appears a characteristic phenomenon: because of the disappearance of the m. latissimus dorsi the attempt mentioned fails and the unfixed shoulder-blades are now partly actively, partly passively and indirectly pushed back and down (by the resistance).

Beside this the appearance of the juvenile dystrophic patient is characterized by dorso-lumbar lordosis and the secondarily protuberant abdomen (Fig. 104). The gait also, because of the atrophy of the glutæi and some of the muscles of the thigh, is waddling and rocking, even though frequently less so than in the infantile forms.

To recapitulate briefly the following muscles are usually *atrophic:* cucullaris, pectoralis major and minor, serratus anticus, latissimus dorsi, the long muscles of the back and loins, somewhat less regularly and later, rhomboidei and triceps, as well as the supinator longus. The following are, as a rule, *spared:* sternocleido-mastoideus, levator anguli scapulæ, coracobrachialis, teres major and minor, supra- and infra-spinatus, the flexors and extensors of the forearm and the small muscles of the hand. In the lower extremities the following are nearly always atrophic: m. quadriceps and the glutæi, somewhat less frequently the flexors of the thigh (m. biceps, semitendinosus, etc.), and the adductors, very rarely the m. tibialis anticus, the peronei and the extensors of the toes. The calf musculature (in respect to function) mostly remains normal, but very frequently it shows signs of pseudo-hyper-

trophy. In the upper half of the body the m. supra- and infra-spinati and the m. triceps are not infrequently hypertrophic.

The mechanical excitability of the atrophic muscles (general and idio-muscular) shows a simple diminution, or it may be lost. Likewise, electrical tests show only a *quantitative* change up to the loss of the reaction; in a few cases, a suggestion of R.D. is said to have been observed. The tendon reflexes usually disappear in the regions that are affected; frequently, also, I have found *general* abolishment of the tendon and periosteal reflexes. The cutaneous reflexes remain intact.

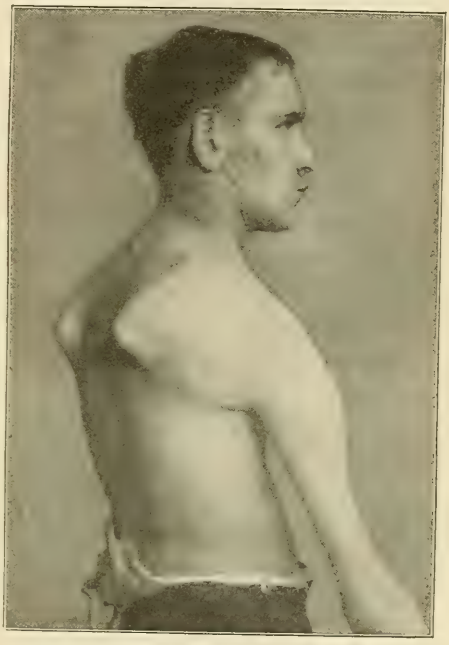


FIG. 103.—Juvenile form of dystrophy. Atrophy of the face (lips), pronounced atrophy of the m. cucullares, serrati (wing position of the shoulder-blades) "sunk in shoulders" and atrophy of the muscles of the upper arm. (*Heidelberg Medical Clinic.*)

Anatomy.—Brain, spinal cord, and peripheral nervous system have (in pure cases) been found normal in all cases examined thus far, even by the most delicate histological methods.

The muscles, on the other hand, always show striking changes: in the atrophic muscles numerous muscular fasciculi are completely destroyed and their spaces filled up by the infiltration of fat; the greater number of the individual muscular fibres are markedly atrophic, but a part of them are also of abnormally large circumference; the muscle nuclei are increased, the muscular fibres themselves usually show fissures and vacuoles, beside proliferation of the interstitial connective tissue. While in pseudo-hypertrophic muscles, although the atrophy, the fatty degeneration and proliferation predominate, still there are muscles which, owing to an extraordinary increase in volume in the original fibres, are to be considered genuinely hypertrophic.

Ætiology.—An actual "exciting" cause of dystrophy is not known. That traumatism could fill this position I consider very doubtful. The disease is to be interpreted rather, as the expression and consequence of a specifically weak congenital predisposition of the muscular system, which succumbs to use, sooner or later, in this typical way.

Course and Prognosis.—The course is always very chronic; whereas the infantile patients rarely reach adult life, dying, as a rule early, of intercurrent diseases; the juvenile form lasts a long time. There are not a few cases in which, dystrophy notwithstanding, the 50th and 60th years was

reached. The progress is usually very slow; especially juvenile cases frequently show a standstill of the disease for ten years and more. The prognosis quoad valetudinem is always bad. I know of only one case through (verbal) communication of *Erb*, in which dystrophy, so diagnosed by him, in a little girl, resulted in recovery.

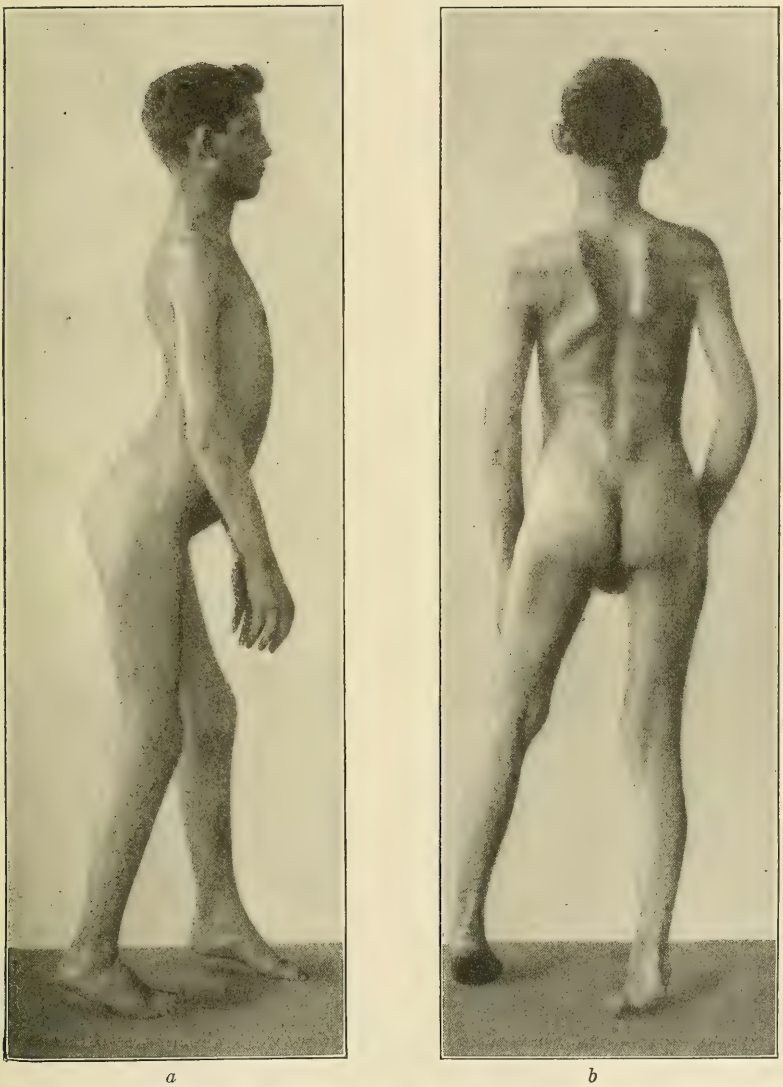
Treatment is correspondingly almost always powerless, even though mild hydriatic, gymnastic and electric procedures are occasionally supposed to have caused improvement (in connection with the rest of the hospital treatment). The organ-therapeutic preparations have proved to be ineffectual. In some cases (especially in the infrequent complications with contractures) orthopedic surgical interference may be indicated. Supporting corsets, on the whole, have not proved very serviceable.

Differential diagnosis must consider in the infantile form, the similarity —which, however, is quite superficial—to diplegia infantum; this, however, may be immediately distinguished by its pyramidal tract symptoms. Poliomyelitis anterior could but rarely be confused with dystrophy. In the pseudohypertrophic form a possible confusion with myotonia congenita must be thought of; however, the characteristic disturbance in motility and muscular reaction of the latter should at any rate easily establish the diagnosis. Differentiation from *Thomsen's* disease with muscular wasting may be more difficult, since the myoatrophy here occasionally shows the typical distribution of dystrophy. But even in such cases the existence of myotonic reaction of individual muscles will decide the differential diagnosis. Furthermore, the rare cases of actual genuine muscular hypertrophy (either congenital or acquired) may appear in differential diagnostic competition, and also the very rare cases of hypertrophy following thrombosis, neuritis and in spastic spinal paralysis. A consideration of the manner of the distribution of the muscular dystrophy will make the diagnosis in all these cases easy. The not uncommon cases of congenital defects of single muscles (especially of the trapezius and pectoral muscles) may also arouse a slight suspicion of dystrophy.

Most important in differential diagnosis are, naturally, the other forms of muscular wasting: the spinal, the neural (*Marie-Hoffmann's*) amyotrophy, syringomyelia, the muscular atrophies in polyneuritis and the already mentioned myotonia amyotrophica. The first two forms mentioned are immediately distinguished from dystrophy, by the fact that they show an opposite arrangement of atrophies: the preference for the *distal* ends of the extremities, whereas dystrophy shows preference for the *proximal* parts. In rare cases, of course, one must reckon with the beginning of the spinal affection in the muscles of the back. The same thing holds true of the polyneuritic forms of muscular wasting. Syringomyelia may indeed, even if rarely, exhibit its earliest atrophy in the proximal arm muscles and the shoulder-girdle; but in this case the disturbance in sensation will immediately decide the differ-

ential diagnosis. But above all, in all the before- mentioned conditions, the appearance of fibrillary tremors and of the electrical reaction of degeneration make the diagnosis considerably easier.

One must mention, finally, that occasionally inflammatory diseases of



a *b*

FIG. 104, *a* and *b*.—Lordosis and typical gait in juvenile dystrophy. (*After Heinr. Curschmann.*)

the muscles with atrophy (*Oppenheim*) and certain diseases of the bones (e. g., rachitis tarda (*author*)) may remind superficially of dystrophy.

Rare Forms and Complications.—Here, first and foremost, the form observed by *J. Hoffmann*, with bulbar paralytic localization of dystrophy, must be mentioned. Also by *Oppenheim* and others, the (very rare) par-

ticipation of the muscles of the tongue and pharynx has been established; likewise the spread of the disturbance to the muscles of the eye. That the dystrophy may begin, quite exceptionally, in the legs proper, in the hands and forearms, was proved by the reports of *J. Hoffmann, Schultze and Erb.* Furthermore those cases are remarkable, in which the trophic changes are not confined to the musculature, but extend also to the bones (*Friedreich, E. Schultze, Schlippe and others*); some of these cases show in addition extensive early contractures in the muscles. Anomalies in the formation of the skull and thorax have also been observed.

We must mention, furthermore, that dystrophy has been observed in combination with all sorts of organic and functional nervous disease (for instance, with tabes, epilepsy, hysteria or psychoses). It is probable from late observations, that there are forms of dystrophy, which never attain complete development (formes frustes).

2. MYOTONIA CONGENITA, THOMSEN'S DISEASE

This rare disease, described first by *Ch. Bell* and *Leyden*, but in the year 1876, actually "discovered" by the Silesian physician *Thomsen* and classically worked up by *Erb* must still be regarded—like progressive dystrophia musculorum—so long as no stringent proof to the contrary be adduced, as a primary myopathy, as a severe intention neurosis of the muscles.

The disease, which in many cases, especially in the family of the discoverer, appears mostly *familial*, with a preference for males, begins in the typical cases usually in early youth, and not seldom, too, about the time of puberty. It usually progresses very slowly, rapidly only upon the occurrence of unusually strong physical exertion (military service) and is then occasionally heightened to an acute onset. The first symptoms are nearly always a certain stiffness and difficulty in walking, rising, lifting, etc., after a period of rest, especially in the morning. This stiffness diminishes upon the repetition of the movement concerned and finally disappears altogether.

The typical picture of the completely developed myotonia, is as follows: the young men, who are usually well-nourished and not seldom full-blooded, show an almost athletic development of the muscles of the trunk and the extremities, which contrasts strikingly with the functional weakness of which they complain. At every first attempt at movement in any group of muscles, after they have been rested (especially in the extremities), there is a peculiar tension and stiffness, a veritable spasm of the muscles concerned, which causes a marked slowing of the movement; and this "myotonus" usually reaches its height at the second movement (not the first); for instance, if the first adduction of the thumb succeeds with relative speed, the following abduction may meet the strongest resistance. This difference between the first and second movement is exhibited very distinctly in rapidly executed powerful movements.

Upon further repetition of the movement concerned, it becomes freer and freer, the tension is soon relaxed and the movement becomes absolutely normal. The disturbance in the gait appears in a most characteristic form: at the beginning slow, short, dragging steps (veritably spastic) are taken with great exertion—after 8–10 m. the walk is freer and becomes completely normal, quick and elastic at the end of a walk of about 15 m. (for instance, a walk through a hospital ward). The disturbance becomes very serious, if the patient is suddenly forced to make a *brusque* and *quick* movement; for instance, to avoid an obstacle that appears in his path, or the like; the legs may then become as stiff as sticks, the patient falls down like a log, and can only rise with difficulty. The myotonic disturbance also, is usually increased by excitement and by cold.

This disturbance may now—in rarer cases—be restricted to definite muscle areas; for instance, to the legs, hands, tongue, etc.; but mostly, it appears generalized, attacking all the muscles of the trunk and limbs, even if not with the same severity. It often disturbs even the motility of the muscles of the face and of mastication, of the muscles of the tongue and of those that close the eyelid, very rarely those of the throat and œsophagus and the external muscles of the eye; it is doubtful whether the musculature of respiration and the heart ever participate.

The affected muscles do not usually show an increase in tonicity while at rest and during passive movements; the passive movements are completely free, in contrast to the active ones; the same is true of the reflex movements (for instance the plantar reflex). On the other hand, the *mechanical and electrical excitability*, as *Erb* first showed, are changed in a most striking way.

The *mechanical excitability* is usually perceptibly augmented; every tapping of the muscle, often mere pressure or kneading, produces an abnormal continuation of the local muscular contraction for from 5 to 30 seconds; there arise, according to the location and form of the muscles, either weals or deep grooves and furrows (with special distinctness in the tongue, the m. gastrocnemius, etc.). At the same time, the idiomuscular excitability is very slight; Schiff's waves are also absent.

The *electrical excitability* is usually absolutely increased for both kinds of currents. With the usual faradic (direct) stimulation, employing somewhat stronger currents, there is occasionally a somewhat slow contraction with long prolongation (2 to 20 seconds or more); single opening shocks of any strength whatever cause only normal, quick contraction; with strong stabile faradization, there appear, occasionally, oscillating muscular waves. The contractions caused by (direct) *galvanization* also are slow, tonic, and show abnormal prolongations; usually only closing contractions (ACC= CCC) appear; in stabile galvanization, occasionally (not always and not in all muscles) rhythmical wave-like contractions from the C to the A. This electric symptom complex is named, according to *Erb*, "*myotonic reaction.*"

The nerve trunks usually do not show any peculiar change in excitability upon mechanical and electrical stimulation; with *indirect* electric stimulation, therefore, there is no myotonus.

The sensory functions, the sphincters, the sensibility and the cutaneous reflexes have always been found normal; the tendon reflexes were occasionally weakened, or abnormally easily wearied, but, as a rule, active. Trophic and vaso-motor disturbances are nearly always absent. The psyche is usually normal—which must be emphasized as against the classification of the disease under the psychoses by *Thomsen* himself—even though a combination with hysteria and epilepsy has been observed.

Varieties of Myotonia.—The most important variety is the one accompanied by different forms of muscular wasting, *amyotrophic myotonia*, which has been observed rather frequently in recent times—in about 20 to 25 cases. In contrast to typical myotonia, it does not appear to begin in early youth, but not until the second to the third decade, to run its course simultaneously with atrophy and pareses and myotonic disturbances. The myotonia may remain more or less generalized, but may also limit itself to only a few muscles (especially to the hands and tongue). The atrophies may be of the type of *Erb's* dystrophy, now but more rarely, of that of the *Duchenne-Aran* amyotrophy (forearm-peroneal type); atrophy of the muscles of the face and mastication is relatively frequent (spontaneous dislocation of the mandibula), as well as speech disturbances, but ptosis (*Fürnrohr*) is very rare. Once inceptive atrophies were proved present in a previously typical case of congenital myotonia (*Hoffmann*). The myotonic reaction is, of course, modified by the atrophies; occasionally one finds the myasthenic reaction. The tendon reflexes are not rarely entirely absent in the amyotrophic myotonia; sometimes there is mechanical over-excitability of the nerves. The course, because of the atrophies, is more severe, but improvements are possible.

The combination of syringomyelia with myotonia, which has been observed in some cases, is also to be placed in the domain of amyotrophic myotonia.

Myotonia acquisita has been described by *Talma* in cases in connection with traumatisms and acute infections. The prognosis in it seems more favorable than in idiopathic cases.

Furthermore, in very rare cases, there have been observed instances of intermittent myotonia; myotonia-like neuroses, accompanied by intermittent paralyses (paramyotonia, *Eulenburg*), of myotonia combined with tetany (*Bettmann, v. Voss*), of myotonia of nursing infants and of those suffering with gastric dilatation.

The **differential diagnosis** of generalized myotonia scarcely ever presents difficulties; at most some cases of pseudo-hypertrophic dystrophia musculorum or rare cases of spastic spinal paralysis may display a superficial

similarity to it; the correct diagnosis, however, is easily recognized by the demonstration of the typical disappearance of the myotonus after repetition of the movement and by the myotonic reaction. The partial myotonia, for instance, of the forearms, may bear a slight resemblance to the chronic tonic form of tetany and to occupational cramps, without, however, causing serious differential diagnostic difficulties. The pseudo-spastic paresis of hysteria may also—very rarely and only superficially—resemble myotonia. In the amyotrophic form, however, the separation from other forms of muscular wasting, sometimes succeeds only after an exact electrical investigation, especially of the non-atrophic musculature.

Morbid Anatomy.—So far, the central nervous system has been found to be free from all changes. On the other hand, the muscles, according to *Erb*, show the following changes: enormous hypertrophy of all the fibres, with most profuse proliferation of the nuclei of the sarcolemma, changes in the finer structure (indistinct striation, formation of vacuoles, etc.), besides a slight increase of the interstitial connective tissue and deposit of a granular substance. *Schiefferdecker* recently proved that the nuclear increase is only relative and that in the sarcoplasma, peculiar grains are found, when a certain fixing agent is employed.

The actual nature of the disease is not yet clear; the heretofore assumed purely myogenous theory is not wholly satisfactory, in spite of *Schiefferdecker's* findings. Several symptoms point perhaps to a central (cerebral?) source of the disease.

Correspondingly, the **ætiology** of myotonia congenita has not been at all explained; the thought of an auto-intoxication (analogy with veratrin poisoning) has only hypothetical interest. The important rôle of heredity speaks for a congenital failure in development of some kind.

The **prognosis** of typical *Thomsen's* disease is quoad valetudinem always bad, quoad vitam always good. The patients are mostly unsuited to vocations which are physically exhausting. The varieties, especially amyotrophic myotonia, the myotonia of gastric dilatation and of babies at the breast have a more serious prognosis.

Treatment is—in typical cases—always fruitless; nerve-stretchings have not proved to be of any value. Careful gymnastics and massage are supposed to be able to induce functional improvements. The main point seems to be prophylaxis with regard to accidents.

3. MYASTHENIA PSEUDOPARALYTICA

(*Myasthenic Bulbar Paralysis, Erb's Disease*)

The first communications concerning this peculiar disease, we owe to *Erb;* later the investigations of *Oppenheim, Hoppe, Goldflam, Strümpell, Eisenlohr, Jolly* and others have taught us more of the nature of the disease and clinically, at least, they have cleared the subject fully.

We must regard the peculiarity of myasthenic paralysis to lie in the fact, that it leads to *serious paralyses of the bulbar nervous realms, first and foremost of the muscles of the eyes and the pharynx, furthermore, to paralytic-like weakness of the musculature of the trunk and the extremities, without ever attaining degenerative atrophies or hypertonic muscular changes, while the macroscopic or microscopic findings in the central nervous system do not show sufficient changes to explain these disturbances.* The paralyses in all the regions have, as *Goldflam* first showed, especially in the beginning, a clear *remittent character;* they are the product of a *morbidly increased capacity for fatigue.* The prognosis is mostly unfavorable; recovery is, on the whole, rare.

Course and Symptomatology.—The disease attacks individuals who have been well heretofore, between the 20th and the 40th years; women are affected somewhat more frequently than men. Usually very gradually, more rarely rapidly after an acute disease, after traumatism or over-exertion, the patients become afflicted with paralytic phenomena of the muscles of the eye, very often with ptosis and diplopia; at the same time one or the other bulbar function, phonation or deglutition suffers. These phenomena may also be associated with a feeling of weakness in the nape of the neck, the trunk and extremities, even though in the beginning the bulbar symptoms—they are often the only symptoms for years—predominate. All disturbances may then disappear for weeks, months or years, without leaving a trace, so that the disease at the beginning may consist of short exacerbations between long remissions. This relatively harmless initial stage may persist for a long time, up to 22 years (*author*). Occasionally, however, the course of the disease is more rapid and approaches its climax in weeks and months gradually, without longer periods of improvement.

It is precisely in the first stage, that this disease shows its characteristic trait, one that distinguishes it totally from all other bulbar symptom-complexes, namely, the *symptom of paralysis, through pathologically increased fatigue:* the muscles of the eye, the muscles of deglutition, which in the morning, after the strengthening rest of the night, perform their functions well or fairly well, fail after a shorter or longer activity, more and more, in the course of the day, until they become paralytic (or more exactly, pseudo-paralytic). A night's rest or some hours of repose during the day suffice to remove the paralysis and the next day the drama of myasthenic fatigue paralysis begins anew. Once the disease reaches its climax or more particularly, if it reaches the final stage, part of the paralyses, however, persist, or show only slight changes according to the condition of rest or fatigue.

Gradually, often too, suddenly, the climax of the disease is reached: the eye muscles, the muscles of mastication and of deglutition, more rarely those of the facial region, frequently the muscles of the nape of the neck and of the back become almost permanently paretic; the fatigue paralysis of the extremities reaches so high a degree, that the patients become permanently

bedridden. Nutrition suffers through the deglutition paralysis; faulty swallowing appears; finally, too, the musculature of respiration fails (possibly also the muscle of the heart) and the end, prepared by inanition, appears quite spontaneously as the result of respiratory or cardiac paralysis, or suddenly after an action that strains the heart (stomach pump, different baths) or is more slowly brought about by an aspiration pneumonia.

Rarely the myasthenia runs its course without actual bulbar paralysis, as pure paresis of the extremities and the trunk (*Grund*, *Rautenberg* and others).

Of the objective symptoms of the completely developed disease the *disturbances of the muscles of the eye* appear most prominently in the fore-

FIG. 105.—Myasthenia with ophthalmoplegia externa on the right side. (*After Knoblauch.*)

FIG. 106.—The same patient trying to look upwards. (*After Knoblauch.*)

ground; the ptosis, which the patients attempt in vain to counteract by auxiliary contraction of the muscles of the forehead, the paralyses of the n. abducentes, of the oculomotor muscles and the rest, so that a total ophthalmoplegia externa results. The defects in mobility frequently seem to be of strictly associated nature (*Bielschowsky*); it is characteristic that the sphincter iridis only rarely is involved in fatigue paralysis. Frequently there exists—in incomplete paralysis of the muscles of the eye—diplopia in a degree that varies from morning to night.

The muscles of mastication become paretic markedly and very early in the disturbance; after a few bites the masseters and temporals give out; swallowing is disturbed by the increasing paralysis of the soft palate, of the muscles of the pharynx and œsophagus; liquid nourishment flows out through

the nose. The laryngeal muscles, mostly the phonatores, rarely then. postici, fail, the voice becomes hoarse, aphonic.

The muscles of the lips, the naso-labial muscles (most rarely the m. frontales), finally almost the entire facial region is paralyzed; the patient has a worried, tired, "mask" face.

At the height of the disease, as we have said, the muscles of the nape of the neck and all the muscles of the trunk show signs of weakness and fatigue; arms and legs, especially in their proximal muscular regions, become tired and paretic after a few movements, after a little lifting and bending, after a few steps.

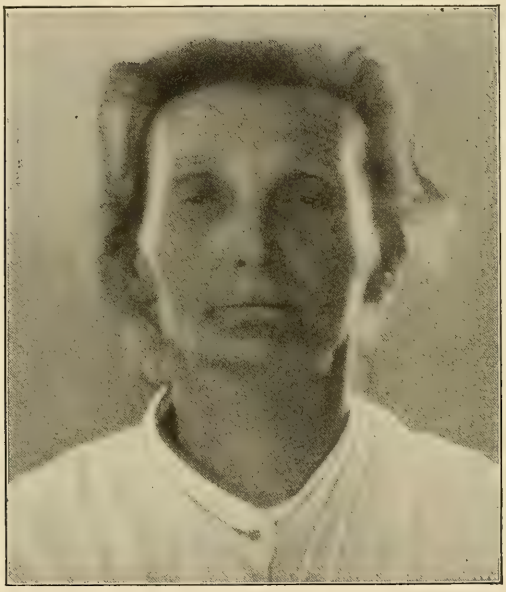


FIG. 107.—Forty-five-year-old patient with myasthenia, bilateral ptosis, myopathic mask-face. (*Author's own observation.*)

At the same time, the paralyzed muscles (in pure cases of the disease) *never present atrophies, fibrillary twitchings, electric or mechanical reaction of degeneration.*

On the other hand, one finds in many muscles (but by no means always in those that are permanently paretic and most seriously affected) *besides the active overcapacity for fatigue, also the same thing for the faradic current, the myasthenic reaction* discovered by *Jolly* (Mya. R.) (Fig. 108); if such a muscle is stimulated by direct tetanization, it reacts at first with normal contractions, gradually the closing contractions lose their intensity and finally fail altogether; after the tetanization has been stopped for a short while (minimum ca. 2 sec.) the muscle is again capable of renewed contraction. The myasthenic reaction does not need to be complete, but it is at least suggested, in

most of the cases. The degree and rapidity with which the Mya. R. is elicited, seems to stand in direct proportion to the former voluntary motor or even electrical fatigue of the muscle.

It must be mentioned that the Mya. R. has been found also in the atrophic muscles in other nervous diseases, in focal injuries of the brain, in "cerebral muscular atrophies" (*H. Steinert*), in amyotrophic myotonia, etc. These findings, however, can not cast doubt on the specific nature of the reaction in the myasthenia, which runs its course without atrophies.

The sensory cerebral nerves (optic, olfactory, glosso-pharyngeal, auditory, etc.) are always *spared* by the disease. The sensory functions of every other sort also always remain intact, usually, too, the bladder and rectum.

The tendon reflexes remain mostly normal; in rare cases they are weakened; often, if frequently elicited, they become overfatigued and disappear. Rarely they may be permanently lost during the period of total paralysis (*author's* observation). The cutaneous reflexes always remain normal.

The psyche of the patient is mostly intact, always, too, the higher functions of speech. Compulsory emotions, the crux of the atrophic and pseudo-bulbar paralytics, seem to be absent.

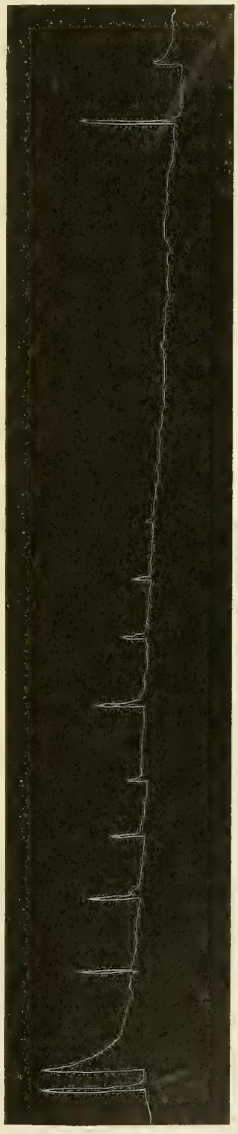


FIG. 108.—Muscle curve in typical myasthenic reaction (m. tibialis anticus). *os*, Opening-closing of the faradic current; *s*, closing of the faradic current; *r*, interruption of faradization. (*After Hedinger.*)

Morbid Anatomy.—The brain, spinal cord and efferent nerves have been found *altogether intact*, in most typical cases, excluding a few in which the changes were insufficient to explain the serious disturbances. On the other hand, in numerous cases, *accumulations of lymphoid cells* have been found in the affected muscles (*Weigert, Flatau, Link, Goldflam, Laqueur, Boldt, Burr, author* and others), which, because of their frequency in the more recent postmortem examinations, possibly have a pathognomonic importance. In one case fatty transformation of the sarcoplasm was found (*Marburg*). In several cases persistent thymus or thymus tumors and muscle metastases (*Weigert*) were found. In one case *Pel* found leucocytosis

intra vitam and leucocytic accumulations in the parenchymatous organs. *Taking all in all, however, the anatomic findings thus far do not explain fully the severe functional disturbances of myasthenia.* Whether the morbid preponderance of the "light muscular fibres," recently found by *Knoblauch*, at the expense of the "red dark ones," in myasthenia and the comparative biological reasons of the author for the pathogenesis of myasthenia, will acquire importance, is as yet by no means sure.

Ætiology.—The real cause is unknown. Acute infectious diseases, however (influenza, traumata, intoxications, over-exertions, etc.), are credited as excitant factors. The latter factor seems especially plausible in an unpublished case of *Erb*, that of an old Boer, who was chased on horseback with Dewet by the English for weeks with minimal resting periods.

Often, however, there is no obvious causal factor to be found.

The assumption that a secretory disturbance of the glandulæ parathyreoideæ is the cause of myasthenia (*Lundborg*, *Chvostek*) has not yet been proven in any way.

Of great importance, however, is the proof first adduced by *Oppenheim*, that many myasthenics exhibit a striking *congenital inhibition in development and deformities* (anomalies in the nervous apparatus, polydactylia in feet and hands —*Oppenheim*—total aplasia of the genital organs and hyperplasia of the lobes of the lungs—observation of the *author*). These findings stamp the patient doubtless as belonging to the congenital hypoplasts, in regard to which *Edinger's* theory of abnormal "using up" and insufficient reparation in the nervous system, must seem very probable to us, and explicative of many things.

The **complications,** by which the disease is rarely accompanied—hysteria, Basedow's disease, dystrophia musculorum, Banti's disease (*Mohr*), can be of little assistance in the ætiological research.

The **differential diagnosis** may be difficult in the beginning, especially relative to hysteria, but is cleared up by the type of the paralysis of the eye muscles, the disturbance in deglutition, the strictly remittent character of the paralyses and finally the Mya. R. Against the diagnosis of atrophic and pseudo-bulbar paralysis we are protected, on the one hand, by observation of the degenerative atrophies, on the other by the disturbances of the lateral pyramidal tracts (spasms, increased reflexes) in those cases. Differentiation from a polyneuritic bulbar paralysis is more difficult in the beginning; the same difficulty is met with in cerebral syphilis, *Landry's* paralysis and especially poliencephalomyelitis (*Oppenheim*). The absence of the quantitative and qualitative electrical changes and the appearance of the Mya. R. will, however, decide the differential diagnosis, even in these cases.

The **prognosis** is usually unfavorable. Although *Erb*, *Goldflam*, *H. Steinert* and others observed recoveries in light cases, most cases after shorter,

or, as usually happens, longer duration, end, as has been described above, fatally.

Treatment can consist only of great conservation of effort, rest and suitable nourishment; besides tonics may be useful. All severe electric or active and passive gymnastic procedures, which could only increase the exaggerated tendency of the muscles to become fatigued, are contra-indicated. Treatment by baths is to be employed only with attentive control of respiration and pulse; better, not at all (*Steinert*). The stomach-tube, which is occasionally unavoidable, when the swallowing apparatus is totally paralyzed, is to be used only with the greatest caution, since cases of death during its passage have been observed (*Oppenheim*).

4. MYOTONIA CONGENITA (Oppenheim)

This disease, first described by *Oppenheim* in 1894, since then described in only about ten cases, seems to be nearly always present at birth, though occasionally—in milder degrees—it was noticed first in the 2nd or 3rd year of life. The children thus affected are conspicuous in that they cannot move their limbs, especially the lower extremities, spontaneously: the limbs are completely paralyzed and flaccid, the muscular tonus is markedly reduced, the limbs hang loosely from the joints. In the arms, the disturbance is usually slighter and here the weakness of the movements is more prominent. The motility of the trunk and neck, on the other hand, is scarcely ever disturbed. Involvement of the muscles of the head, face, tongue, eyes, larynx and diaphragm has not yet been observed. The sensory cerebral nerves, the sensory functions, the sphincters and the intellectual development, also, seem not to suffer in the disease.

The muscles, though they feel limp and withered, show *no real atrophies* and no fibrillary twitchings. The direct and indirect electrical excitability in the paralyzed as well as in the apparently sound muscles is usually reduced, *quantitatively* to a large degree, frequently even to the point of entire disappearance of the excitability, and a suggestion of the reaction of degeneration has been observed.

The tendon reflexes are nearly always abolished, not only in the extremities affected, but also in those that are normally movable; the cutaneous reflexes are mostly described as normal.

Anatomic findings are known so far, only in one case (*Spiller*): the entire nervous system proved to be intact; the muscles, on the other hand, showed hyaloid changes, looked cloudy and had only indistinct striation; the individual fibrils were abnormally small, the fat containing connective tissue increased. Signs of inflammation and breaking down were lacking. Other authors corroborated these findings in sections of muscle taken from the living body (*Reyher and Helmholtz*), whereas *Bing* found normal structure.

Oppenheim sees the very *essence of myatonia* in an inborn retarded development of the musculature; the anatomical findings for the most part, seem to speak for this. *Bernhardt* is rather inclined to assume an injury to the peripheral nerves, somewhat like a generalized polyneuritis of autotoxic or infectious origin (partly on the basis of cases, which were not congenitally diseased). At any rate, the pathogenesis of the disease is greatly in need of classification by further clinical and anatomical findings, which first and foremost must prove if there is really such a disease entity.

The **differential diagnosis** must consider especially, poliomyelitis anterior and the peripherally and spinally induced paralyses intra-partum. The absence of atrophies and disturbances in sensation, the generalized, almost always symmetrical affection of the muscles and the change in electrical excitability, which mostly affects *all* the muscles, distinguishes, however, myatonia from those diseases. From a—probably exceedingly rare—general polyneuritis of nursing infants, the disease, it seems to me, can not be distinguished. Finally, I desire to earnestly point out, that nervously normal, but somewhat atrophic or rachitic children may show a high degree of hypotonia of the muscles and joints (which, to a certain degree, is physiologically peculiar to early childhood). This likewise furnishes a differential diagnostic source of error.

The **course** is always extremely slow. The *prognosis*, however, is not bad, since in some of the cases, in the course of time, a more or less complete disappearance of the disturbances has been observed.

Treatment must consist in the use of general tonic means, of careful strychnine medication, massage and the electric current; possibly orthopedic procedures may also become necessary.

V

DISEASES OF THE BRAIN

NORMAL AND PATHOLOGICAL PHYSIOLOGY OF THE BRAIN[1]

BY

HUGO LIEPMANN (Berlin)

I. INTRODUCTORY REMARKS ON THE ANATOMY, PHYSIOLOGY, AND PATHOLOGY OF THE BRAIN

1. The Membranes of the Brain

The brain, covered by two membranes, the dura mater and pia-arachnoid, lies enclosed within the cranium.

The topographical relations of the different parts of the brain to the skull are shown in Fig. 109.

The dura mater is the periosteum of the bones of the skull. In the healthy adult it is attached firmly only to the base of the skull, and can be easily detached from the convexity. Extensions of it project sagitally into the spaces between the hemispheres of the cerebrum (proc. falcif. major.), the hemispheres of the cerebellum (proc. falcif. minor.), and transversely between the cerebellum and the base of the occipital lobe (tentorium).

It forms, in places, by splitting into two layers, the so-called sinuses. They contain blood. Its fibrous layer forms sheaths for the cranial nerves as they pass through the foramina.

Its nervous supply arises partly from the sympathetic and partly from the trigeminal.

It is impossible to give the varying views of the leading specialists on the many mooted points.

The names of authors are purposely not quoted, except in a few cases, in which a fundamental theory is connected with a name. The soft membrane is divisible into two, an outer layer, the *arachnoid*, and an inner, the *pia mater*.

The pia lines all fissures, following closely the surface of the brain and enters the ventricles as the tela chorioidea, while the arachnoid membrane is stretched across the fissures. The two are closely connected by numerous trabeculæ of connective tissue, so that pathologically, they may be treated

[1] The need of brevity prevents minute discussion and compels us to present a very intricate matter in as simple a form as possible.

as one. Between the dura and the pia arachnoid lies the *subdural space*, which contains scarcely a trace of fluid. Even in disease it is not a place for the collection of fluids.

On the other hand in the subarachnoid space, between the arachnoid membrane and the pia, because of its communication with the ventricles, is found normally some cerebro-spinal fluid, which, in progressive paralysis, senile atrophy, etc., is often much increased (hydrocephalus externus).

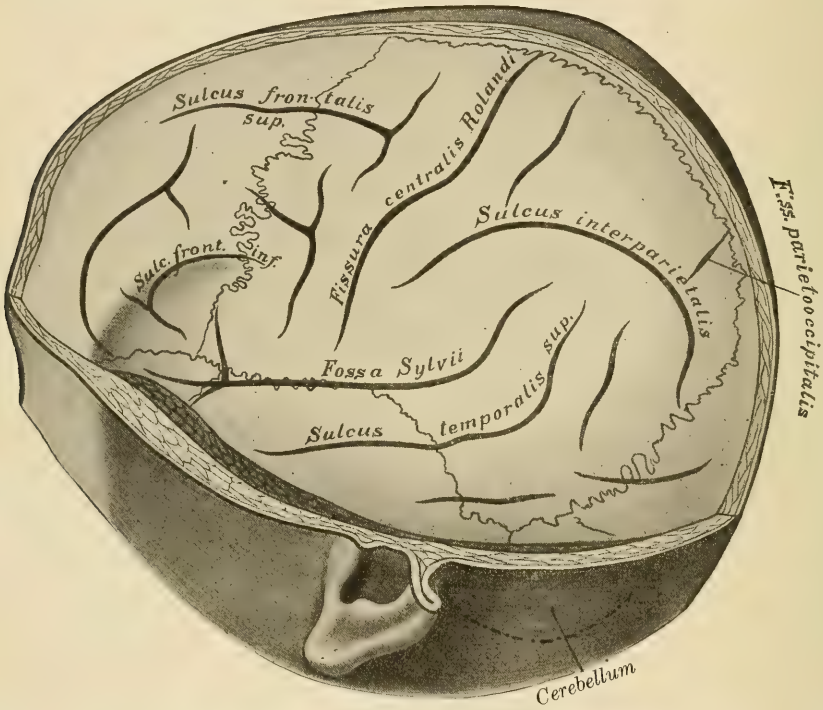


FIG. 109.—Relation between the cranium and the cerebral surface. (*After Bardeleben-Haeckel.*)

2. Circulation in the Brain

The brain receives blood from

1. Both vertebral arteries.
2. Both internal carotid arteries.

Both vertebral arteries unite in passing from the medulla oblongata to the pons and form the basilar artery, which before reaching the pons divides into the two posterior cerebral arteries coursing backward and laterally. —Art. cerebr. post.

The internal carotid branches as follows on both sides:

(a) Anterior cerebral artery (Art. cerebr. ant.).

These two are connected by the anterior communicating artery.

(b) The middle cerebral artery (Art. foss. Sylvii).
(c) The posterior communicating artery (Art. commun. post.).
(d) The chorioidal artery (Art. chorioidea).

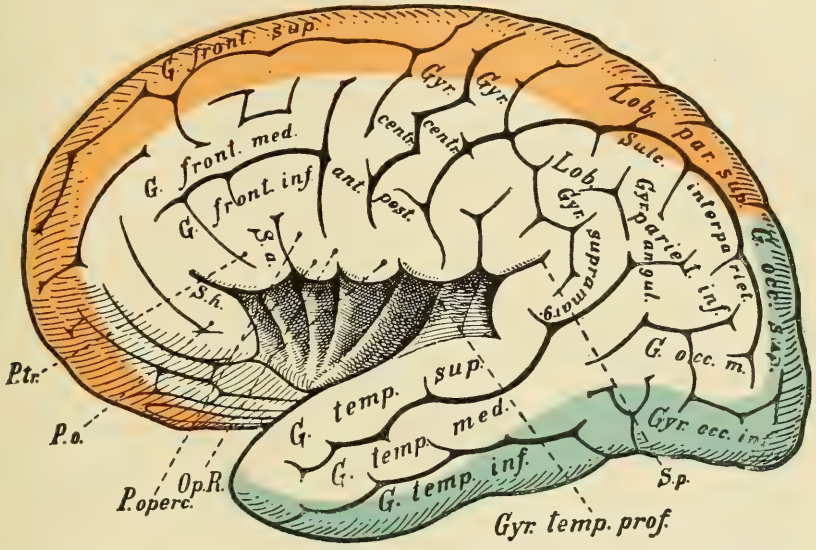


FIG. 110.—Areas of the convexity as supplied by the cerebral arteries. White, area of the art. fossæ Sylvii. Red, of the art. cerebri anterior. Blue, of the art. cerebri posterior. The other abbreviations as in Fig. 112.

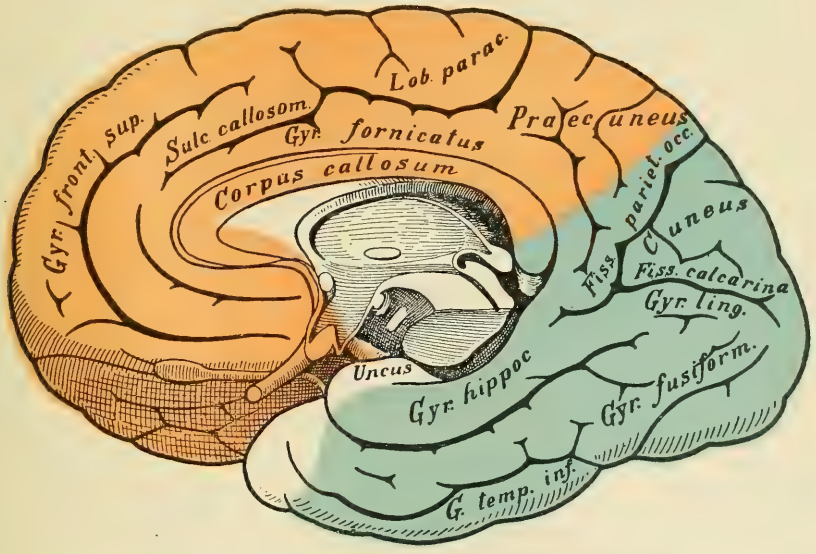


FIG. 111.—Areas of the median surface as supplied by the cerebral arteries. Colors as in Fig. 110.

In the posterior communicating artery is mixed the blood from the carotid with that from the vertebral, as it joins the posterior cerebral artery. ⌜By

means of the anterior and posterior communicating arteries the circle of Willis is completed. Variations in it are frequent.

The three main cerebral arteries are therefore:

1. The anterior cerebral artery. } (Both from the carotid.)
2. The middle cerebral artery. }
3. The posterior cerebral artery (from the basilar artery).

These main arteries send *short* branches into the inner part of the brain (central arteries), especially to the ganglia, and *long* branches to the cortex, and the adjacent white matter (cortical arteries).

The short and long (cortical) branches form, in themselves, two complete systems of circulation, not interdependent but independent of each other, as there are no anastomoses between them.

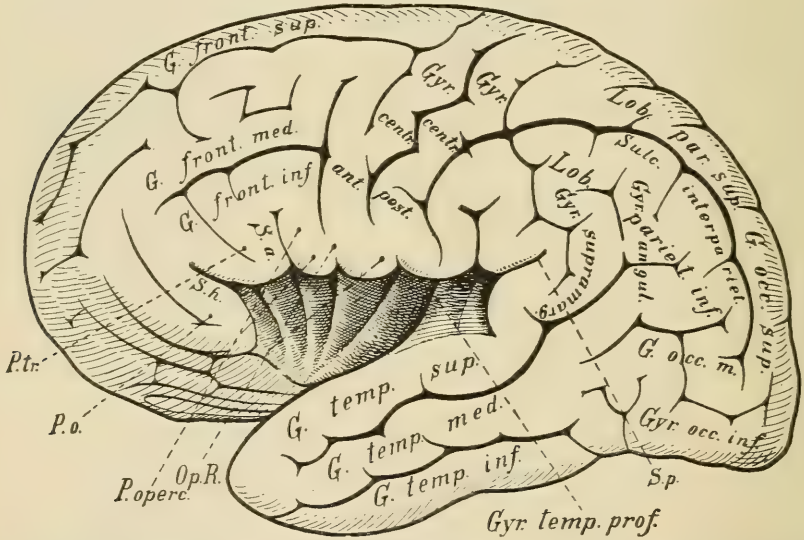


FIG. 112.—Convolutions and fissures of the convexity. (Using a design of Flechsig.) *S.h.*, *S.a.*, *S.p.* Ramus horizontalis, ramus ascendens or verticalis and ramus posterior of the Sylvian fissure; *Op.R.*, operculum Rolandi; *p.tr.*, pars triangularis of the third frontal convolution; *P. operc.*, pars opercularis of the third frontal convolution; *P.o.*, pars orbitalis of the third frontal convolution. The designation *Gyr. supramargin* ought to begin more in the front immediately behind the *Gyr. centr. post.*

The short branches (central arteries) have among themselves as well as with other arteries few anastomoses, so that they are designated terminal arteries.

The long branches course along on the surface of the brain, supply the cortex and adjacent white matter, and have, among themselves, anastomoses.

Of the central branches, those that supply the inner capsule, the lenticular nucleus and the optic thalamus, are of especial importance. (Lenticulostriate and lenticulo-optic arteries). The region supplied by these non-anastomosing vessels, is the *place of preference*, for the bursting of blood-vessels and cerebral softening.

The *anterior cerebral artery* (main branch: the corpus callosum artery: art. corp. callosi) supplies both superior frontal convolutions, the base of the frontal lobes and the medial parts of the central hemisphere (gyrus fornicatus, the paracentral lobule including the præcuneus, and the corpus callosum).

Since the paracentral lobule is the seat of the leg-center, an obstruction in this branch of the anterior cerebral artery produces an *isolated paralysis of the leg.*

Disease of the *middle cerebral artery* or the artery of the fissure of Sylvius has especial importance in disturbances of speech. Its short central branches have already been mentioned, as the important lenticulo-striate, etc.

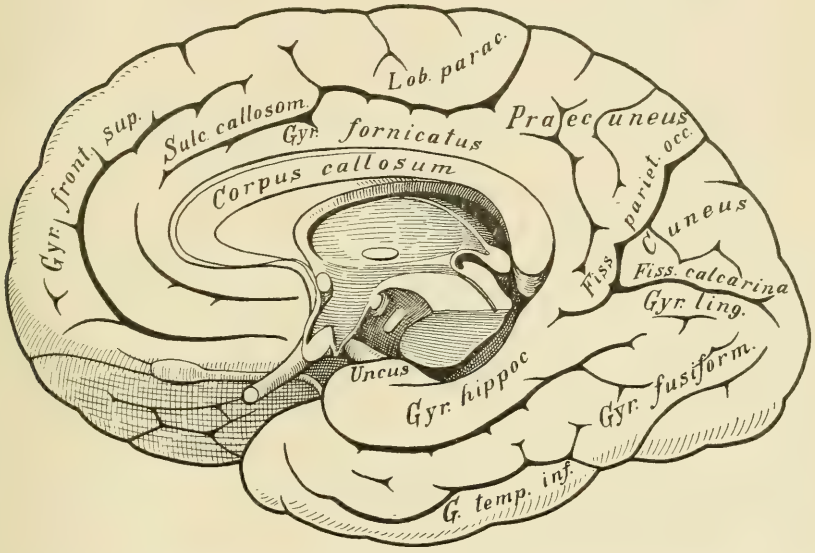


FIG. 113.—Convolutions and fissures of the medial surface. (Using a design of Flechsig.) *Sulc. callosom.*, Sulcus callosomarginalis; *Lobus parac.*, lobus paracentralis.

Its 5 to 7 main cortical branches supply the third frontal convolution the island of Reil, the central convolutions, the parietal lobes, and the superior convolutions of both temporal lobes.

The *posterior cerebral artery* supplies the walls of the ventricles. Its short branches go to the optic thalamus, the crus cerebri, the geniculate body, the chorioid tela, etc. By short branches it supplies also the splenium of the corpus callosum.

It divides into two main cortical branches:

1. The *temporal artery* which supplies the undersurface of the temporal lobe.

2. The *occipital artery* which supplies the occipital lobe.

Of the branches of the latter the calcarine artery is especially important, because it supplies the vicinity of the calcarine fissure—that is the visual center—and the optic radiation.

The *cerebellum and the pons* are supplied by branches of the basilar artery (the cerebellar artery).

The different parts of the brain are, therefore, supplied as follows:

The *cortex of the cerebrum and its white matter;* by the *cortical* branches of the three main arteries, thus:

The *frontal brain:* basal, medial and upper convexity, by the anterior cerebral artery; the rest of its convexity, especially F. m. and F. i. by the middle cerebral artery.

The *central convolutions, the island of Reil and the convexity of the parietal lobes* by the middle cerebral artery.

The medial parietal lobes (paracentral lobule and præcuneus) gyrus fornicatus, by the anterior cerebral artery. (Præcuneus partly also by the posterior cerebral artery.)

The temporal lobes, the two upper convolutions by the middle cerebral artery; the rest, especially the base, by the posterior cerebral artery.

The occipital lobes, by the posterior cerebral arteries.

The large gangliæ and inner capsule, by the *central* branches of the three main arteries.

The corpus callosum by the anterior cerebral artery; the splenium by the posterior cerebral artery.

The middle basal region, by the posterior communicating artery.

The cerebral peduncles, by the posterior cerebral and the posterior communicating arteries.

The quadrigeminal bodies, by the posterior cerebral artery.

The pons, cerebellum and *medulla oblongata,* by the superior cerebellar artery, the basilar artery and the inferior cerebellar artery, the vertebral artery.

Veins of the Brain

The veins of the brain have no valves and are connected by innumerable anastomoses. They drain into the dural sinuses (sinus longitudinalis, etc.), which finally empty their blood into the internal jugular vein. The veins from the corpus striatum and the choroid plexuses of the ventricles, unite in the azygos great cerebral vein (*vena magna Galeni*) below the corpus callosum. This empties its blood into the straight sinus (sinus rectus).

3. Morphology of the Brain

The weight of the brain in man averages about 49 ounces, in woman 4 ounces less.

On the grounds of embryological development one distinguishes:

1. *The end brain* (telencephalon); both hemispheres with the anterior ganglia; caudate nucleus, lenticular nucleus, amygdaloid nucleus and claustrum. Its cavity: the lateral ventricles.

2. *The interbrain* (diencephalon): the optic thalamus with its appendages, especially the important geniculate body and, at the base, the chiasma and optic tract. Its cavity: the third ventricle.

These together constitute the *forebrain* (prosencephalon).

3. *The midbrain* (mesencephalon): *the quadrigeminal bodies*, inferior lemniscus, red nucleus and the nuclei of the third and fourth nerves, *cerebral peduncles* (tegmentum and base) with the substantia nigra. Of the cerebral cavities the aqueduct of Sylvius corresponds to the midbrain.

4. *The hind-brain* (metencephalon or rhombencephalon): the pons and cerebellum.

5. *The after-brain* (myelencephalon): the medulla oblongata.

The cavity of the hind and after brain is the fourth ventricle. The end towards the spinal cord forms the lowest part of the pyramidal crossing.

The forebrain, together with the midbrain is frequently spoken of as the *greater brain* (cerebrum) as against the cerebellum, the medulla and the pons.

The "*brain stem*" is the name for the medulla, pons, midbrain, interbrain and the cerebral ganglia in contrast to the cortex and the white matter of the hemispheres.

Each hemisphere is divided into frontal, parietal, temporal and occipital lobes.

The largest fissure, the *fissure of Sylvius*, separates the temporal lobe from the frontal and parietal lobes. Many claim that the frontal lobe is separated from the parietal lobe by the fissure of Rolando or central fissure, so that the anterior central convolution is counted as belonging to the frontal, the posterior central convolution as belonging to the parietal lobe. But both central convolutions are often spoken of as a whole: the *central region* or *region of Rolando*, and in that case it counts as an independent part lying between the frontal and parietal lobes.

Between the frontal, parietal, and temporal lobes, in the depths of the fissure of Sylvius lies the *island of Reil* (*insula*). The parts of the frontal brain and of the central convolutions covering it are, hence, called the operculum, and that part of the operculum that belongs to the central convolutions, the *Rolandic operculum*, that, which belongs to the inferior frontal cortex, the *frontal operculum* (Broca's area).

The occipital lobe is divided sharply from the parietal lobe, only on the medial surface, by the parieto-occipital fissure; a distinct boundary is lacking at its convexity.

(a) Convolutions and Fissures of the Convexity.

The surface of the *frontal lobe* is divided by two fissures into three convolutions, the superior or first, F s, the middle or second, F m, and the inferior or third, F i, frontal convolution.

The inferior frontal convolution is divided by the anterior (horizontal) and the ascending (vertical) branch, (ramus ascendens) of the fissure of Sylvius into three parts (Fig. 110):

1. In front, pars orbitalis,

2. Between the horizontal and the ascending branch, the pars triangularis and

3. Behind the ascending branch, the before mentioned frontal operculum which posteriorly gradually disappears in the Rolandic operculum.

The *parietal lobe* extends anteriorly, if we consider both central convolutions as the Rolandic region per se, up to the fissure that marks the back boundary of the posterior central convolution and is divided by the interparietal fissure into a superior and an inferior lobule.

The inferior parietal lobule is divided into

1. The anterior part or the *supramarginal* gyrus which surrounds the posterior end of the fissure of Sylvius and, below, forms the extension of the superior temporal convolution posteriorly, and which extends anteriorly to the posterior central convolution (the French designate only that part of the convolution that touches the posterior end of the fissure of Sylvius, as the gyrus supramarginalis).

2. The posterior part, or the *angular* gyrus (pli courbe) which surrounds the posterior end of the temporal fissure. Between the supramarginal and angular gyri, the inconstant fissure of Jensen.

This angular gyrus, often interrupted by a fissure, continues into the occipital lobe.

To simplify orientation one must remember that the supramarginal gyrus is placed at the posterior end of the fissure of Sylvius, and the angular gyrus at the posterior end of the superior temporal fissure.

The convexity of the *temporal lobe* is divided, by the above-mentioned superior temporal fissure (sulcus temp. sup.) and the middle temporal fissure, into three convolutions, the superior, middle, and inferior temporal convolutions. The superior temporal convolution continues posteriorly into the parietal lobe, especially the supramarginal gyrus corresponding to the end of the fissure of Sylvius, the middle temporal convolution, especially into the angular gyrus and the inferior, into the corresponding occipital convolutions.

Coursing along the surface of that part of the temporal lobe, that is turned toward the island of Reil—concealed in the fissure of Sylvius—taking their origin in the surface of the superior temporal convolution, 1 or 2 *cross convolutions* (*gyri temporales profundi* or *transversi*, also called the convolutions of Heschl), pass backwards and inwards to the parietal region behind the island of Reil.

The *occipital lobe* has anteriorly no sharp boundary. As an artificial boundary one assumes a line that unites the upper end of the parieto-occipital fissure with the incisura præoccipitalis.

(b) The Median Surface and Base of the Brain

The median surface of the hemispheres shows first of all the entrance of the corpus callosum, that great commissural system, which connects both the symmetrical and the asymmetrical parts of the two hemispheres.

Below the corpus callosum, and bent from before backwards in a similar curve, we see the fornix (the thickened ends of the hemispheres), which continues towards the back in the fimbria of the inferior horn of the lateral ventricle (Fig. 113).

Parallel to, and above the corpus callosum, lies one of the main furrows of the median surface, the sulcus cinguli or callosomarginal sulcus, which turns posteriorly upwards to the edge of the hemisphere and ends in a small notch behind the end of the posterior central convolution.

The concentric convolution between this furrow and the corpus callosum, is called the gyrus *fornicatus*. What lies in front of, and above it, is classed with the frontal brain (gyrus rectus and medial part of the superior frontal convolution); only the hindmost part of this concentric convolution, which corresponds to the central convolutions of the convex surface, is considered as a small lobe, per se, called the paracentral lobule (lobus *paracentralis*).

The posterior part of the gyrus fornicatus (back of the ascending branch of the sulcus cinguli), spreads into the *præcuneus*, which is part of the parietal lobe. Its posterior boundary is the second main fissure of the median surface: the parieto-occipital fissure, mentioned before as the boundary between the parietal and the occipital lobes. Into this fissure, anteriorly, passes a third *very important* main fissure, the calcarine fissure (*fissura calcarina*), running sagitally postero-anteriorly.

The wedge-shaped part of the convolution, lying between the fissures, is the *cuneus*.

Under the calcarine fissure lies the *gyrus lingualis*, and under this, separated from it by the occipito-temporal sulcus (collateralis) *the gyrus fusiformis*, which continues anteriorly through the temporal lobe. Both, the gyrus lingualis and fusiformis belong to the basal surface. The gyrus fornicatus continues backwards and upwards into the præcuneus. Towards the lower and back part, it winds about the splenium of the corpus callosum, and here continues into the *gyrus hippocampi*, which extends to the anterior end of the median temporal lobe. The hook-like bend, which the concentric convolutions (gyrus fornicatus and gyrus hippocampi) show at their anterior extremity, is called the uncus.

In the gyrus hippocampus, the gyrus lingualis continues from the back, so that in the gyrus hippocampus the gyrus fornicatus and the gyrus lingualis are united.

As is evident, the median surface, posteriorly, gradually passes into the basal surface.

On the basal surface, between the temporal and the occipital lobes, no sharp line can be drawn. The inferior temporal convolution helps to form the base.

Medially from it, separated by the inferior temporal fissure, lies the before mentioned fusiform gyrus, and medially in front of this, lies the hippocampal gyrus, in back of it the lingual gyrus. The fusiform gyrus is separated from them by the occipito-temporal fissure (collateral fissure)

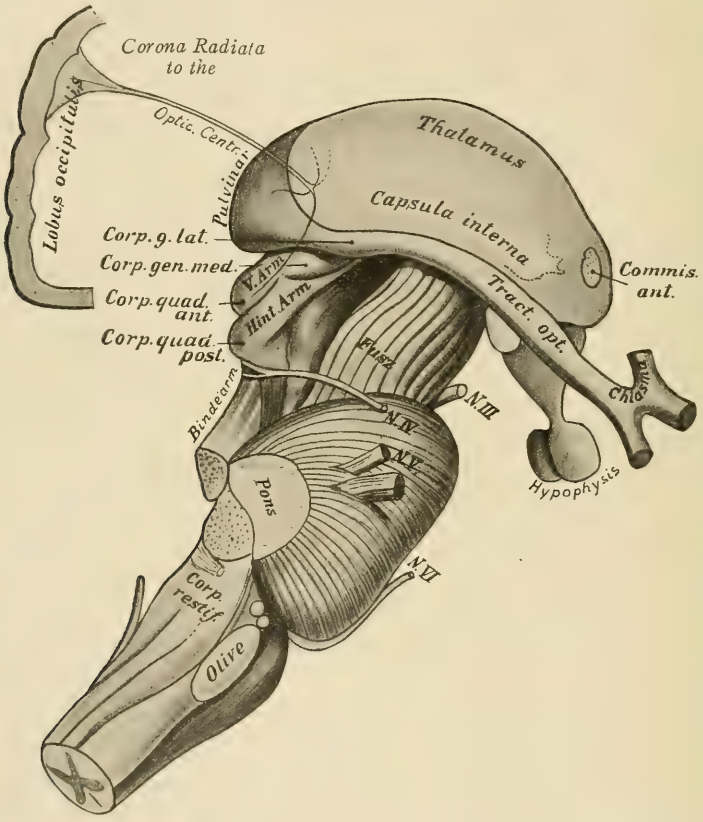


FIG. 114.—Lateral view of inter- and midbrain. The connections between corp. geniculat. laterale (*Corp. g. lat.*) and the occipital lobe, not indicated. (*After Edinger.*)

which extends through the entire undersurface, almost to the temporal pole. In front the base shows between the central peduncles, the mammillary bodies, the pituitary body, the optic tracts, the anterior and posterior perforated spaces.

At the base of the frontal lobe the gyrus rectus, i. e., the basal continuation of the superior frontal convolution is cut off towards the inner side, by the olfactory sulcus in which the olfactory lobe is situated. Outwards from it, lies the basal part of the middle frontal convolution, and separated from it by the sulcus orbitalis, the basal (also called orbital) part of the inferior

frontal convolution (pars orbitalis of F. i.). To understand the form relations of the inter-, mid-, after-, and hind-brain, one needs only to look at Figs. 114 and 115.

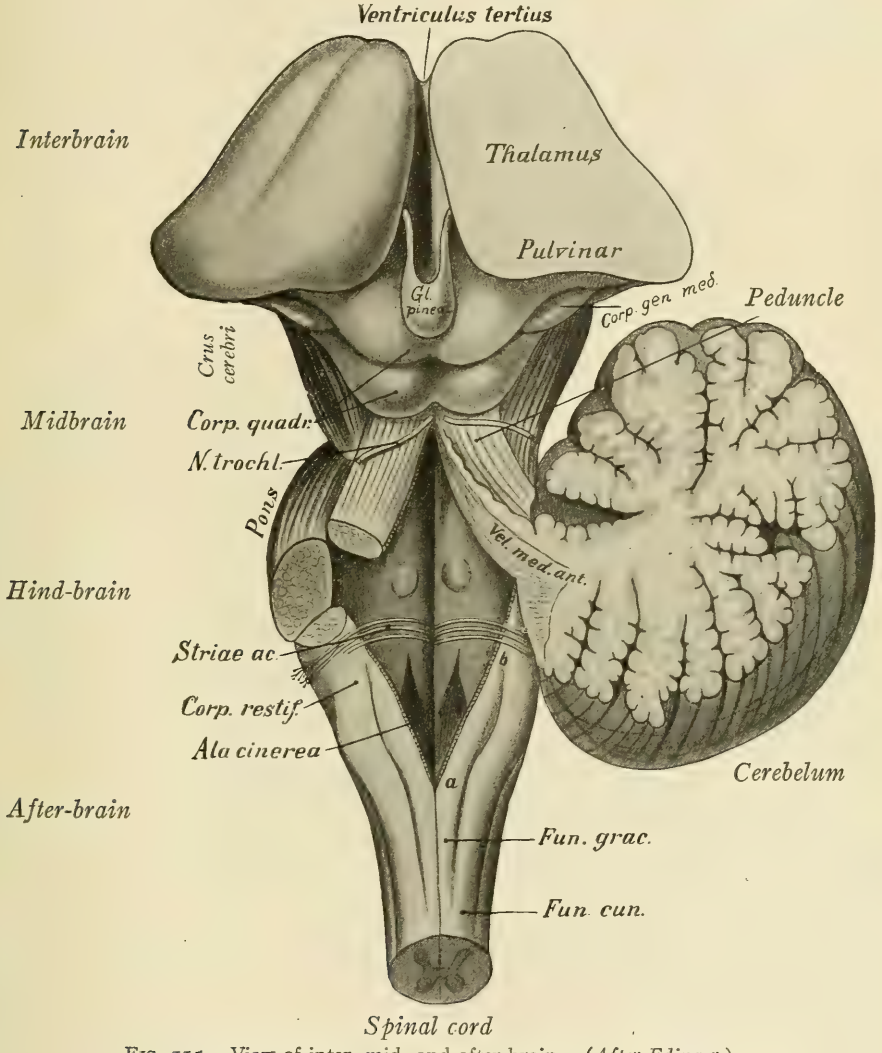


FIG. 115.—View of inter- mid- and after-brain. (*After Edinger.*)

4. Histological Structure of the Cortex

The cellular cortical structure, as proven by embryology and comparative anatomy, may be divided into *six* layers:

1. Lamina zonalis (molecular layer).
2. Lamina granularis externa (outer granular layer).
3. Lamina pyramidalis (pyramid layer).
4. Lamina granularis interna (inner granular layer).

5. Lamina ganglionaris (ganglia cells layer).

6. Lamina multiformis (polymorphous or spindle celled layer).

This six-layered type, established on embryologic grounds, in the *whole* cortex of the cerebrum, is, besides, in Fig. 116 divided into subdivisions (*III a* and *b*, *VI a* and *b*), and is subject to numerous, local modifications which frequently are sharply limited; they consist in increase, diminution, or

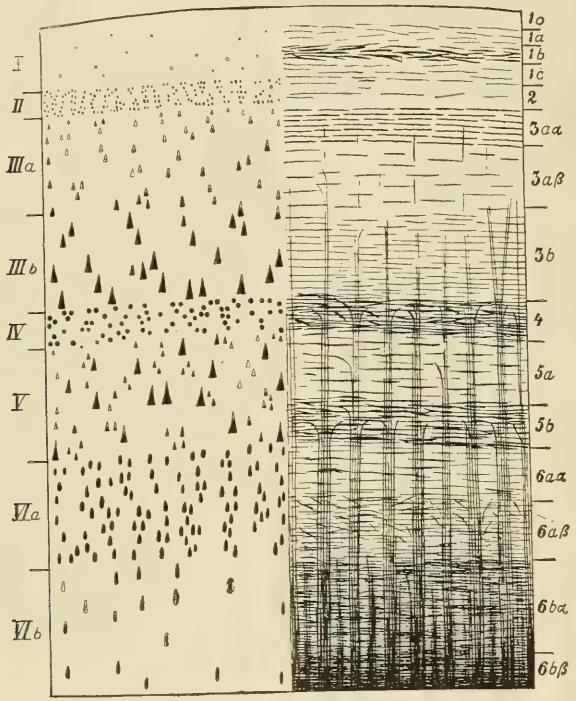


FIG. 116.—Cyto- and myelo-architectonic structure of the cortex. The Arabic figures at the right designate the myelo-architectonic layers. 1*o*, Pars afibrosa laminæ tangentalis; 1*a*, pars externa laminæ tangentalis; 1*b*, pars intermedia laminæ tangentalis; 1*c*, pars interna laminæ tangentalis; 2, lamina dysfibrosa; 3*aα*, stria Kæsi-Bechterewi; 3*aβ*, regio typica partis superficialis laminæ suprastriatæ; 3*b*, pars profunda laminæ suprastriatæ; 4, stria Baillargeri externa; 5*a*, lamina intrastriata; 5*b*, stria Baillargeri interna; 6*aβ*, lamina substriata; 6*aβ*, lamina limitans externa; 6*bα*, lamina limitans interna; 6*bβ* substantia alba. The Roman figures at the left designate the cyto-architectonic layers. *I* to *VI* see text. (*After Vogt and Brodmann.*)

transposition of single layers, and the inner granular layer may disappear, or by fission, double, and other cellular formations, i. e., *giant* pyramid cells may appear and finally variations in the thickness and size of the cells, in their density, and the breadth of the layers, are found. *Accordingly, in the entire cortex about* 50 *such modifications of the layers can be distinguished, and corresponding regions marked off.* In general, their boundaries do not correspond to the fissures and convolutions (Fig. 117).

Of special importance are two extreme variations of cell layers:

1. *The giant pyramidal* or *motor* type in the *anterior central convolution* (Fig. 118). The fourth layer is wanting.

2. *The calcarine type* with duplication of the inner granular layer in the immediate vicinity of the calcarine fissure (corresponding to the *clinico-pathologically* defined visual sphere of *Henschen*).

The regions as mapped off according to the cortical cell architecture (cyto-architectonically), correspond only partly, and only approximately to the regions mapped out by *Flechsig*, on the basis of myelin-development (myelogenesis).

Beside the *cellular* layer, differences in density, arrangement, and caliber of the medullated *nerve fibres* of the cortex, condition a different cross-cut

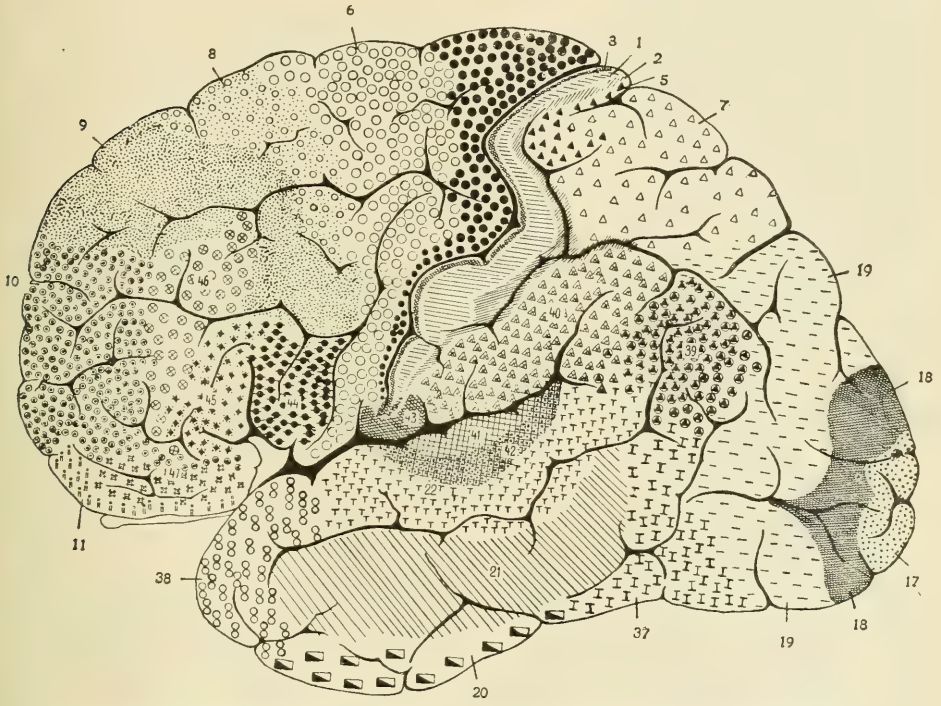


FIG. 117.—The cyto-architectonic cortical regions of the convexity. (*After Brodman.*)

division of various places of the cortex, which gives boundaries of about the same cortical fields, as cyto-architectonics (Fig. 116).

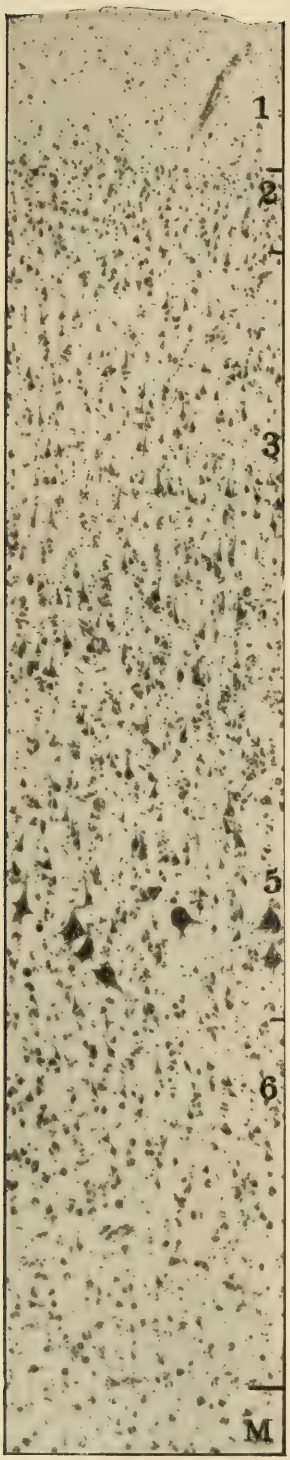
The relation of the *horizontal* fibrillation of the cortex, to the *radial* medullated nerve fibres, if we overlook local peculiarities, presents the following appearance:

1. Tangential layer.

2. Supraradial felt work (*Edinger*).

3. Intraradial felt work, containing the internal and external lines of Baillarger, which form only a density in the fibres that runs horizontally through the cortex.

More detailed investigation has led to a further separation into divisions



and subdivisions, which we, in the above scheme of *O. Vogt* (Fig. 116) have brought into relation with the cyto-architectonic fundamental type of Brodmann.

The peculiarly strong marking of this horizontal fibre layer in the calcarine region is known as the *Vicq d'Azyr* stripe; it is easily seen in a freshly cut brain, appearing as a white line in the cortex.

5. Nuclei of the Cerebral Nerves

1. The olfactory nerve (Nerve I) cf. page 466.

2. The optic nerve (Nerve II) cf. visual paths (Fig. 119).

3. Nerves of the eye muscles: the oculomotor nerve (Nerve III), the trochlear nerve (Nerve IV), the abducens nerve (Nerve VI) are all purely motor.

The nucleus of the *oculomotor nerve* lies in the region of the anterior quadrigeminal bodies, in the floor of the aqueduct of Sylvius. It has a medial division, lying exactly in the middle line, which sends fibres to both sides, and a larger lateral one, on each side, which also sends off fibres to the opposite side. The nucleus consists of aggregations of cells, each of which, probably, supplies a definite muscle. The roots issue from the inner edge of the cerebral peduncle, in front of the pons, the fibres pierce the dura mater, run into the wall of the cavernous sinus, and, through the superior orbital fissure, reach the eye. The nerve is divided into a superior and an inferior division (ramus superior and inferior). From this latter a short branch, the radix brevis ganglii ciliaris, goes to the ciliary ganglion (Fig. 120).

4. The nucleus of the *trochlear nerve* lies in the caudal extension of the oculomotor nerve nucleus in the region of the posterior quadrigeminal

FIG. 118.—The giant pyramidal type in the anterior central convolution. (*After Brodman.*)

bodies. The fibres pass from the nucleus dorsally and *decussate* completely in the velum medullare anticum. They appear back of the quadrigeminal bodies, pass around the cerebral peduncles towards the front and ventrally, and reach, through the wall of the cavernous sinus and the superior orbital fissure (Fissura orbitalis superior), the superior oblique muscle. (Musculus *obliquus* superior.)

5. *The Abducens Nerve.*—The nucleus lies in the region of the pons, in the floor of the fourth ventricle. The fibres pass ventrally, emerging between the pons and the pyramid. The nerve passes through the cavernous sinus and the superior orbital fissure to the external rectus muscle. (Musculus rectus externus.) (Fig. 121.)

Connections between the muscles of the eye, as well as with the Deiter nucleus (of the vestibular nerve) and the cerebellum are made by the *posterior longitudinal bundle* (Fascicul. longitud. post.), which can be traced from the

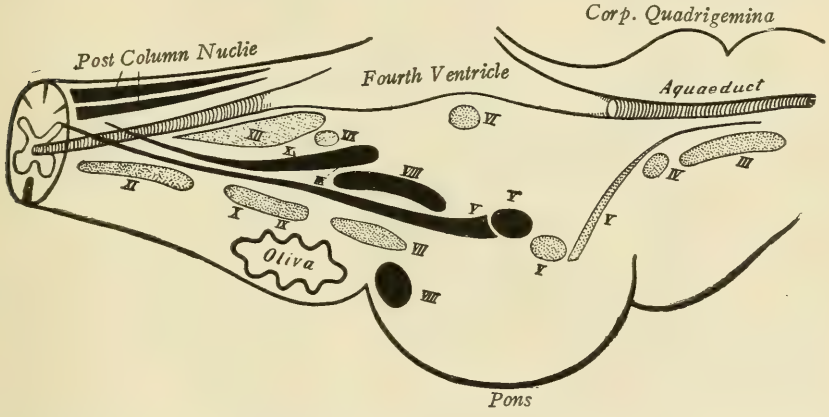


FIG. 119.—Schematic view of the location of the nuclei *Nr. III–XII.* Motor nuclei dotted, sensory deep black. *XIX* means *X* and *IX* (common nucleus of *Nr. IX* and *X*). (*After Villiger.*)

posterior commissure to the spinal cord, and which among other things produces the relations existing between ocular movements and changes in equilibrium.

6. The *trigeminal nerve* (Nerve V), a mixed nerve, has a very large nucleus. The main motor nucleus lies in the dorsolateral tegmental part of the pons. In addition, there is a smaller motor nucleus in the quadrigeminal region at the side of the aqueduct. The motor root passes directly to the third branch of the mandibular nerve.

The thicker *sensory* root has its origin in the Gasserian ganglion, enters the pons, and runs near the sensory end nucleus, where the fibres divide into ascending and descending branches.

The ascending branch ends in a collection of cells, situated next to the motor nucleus in the tegmental part of the pons (nucleus sensibilis N. V);

the descending branch ends in a cell column which is the caudal extension of the before mentioned sensory nucleus. This collection of descending fibres is called the *descending* or *spinal* trigeminal root; it can be traced to the second cervical segment. The caudal part of this sensory nucleus is identical with the gelatinous substance of Rolando in the posterior horn of the spinal cord.

The *facial nerve*, a motor and secretory nerve (N. VII). The nucleus lies in the tegmental part of the pons, ventrolaterally from the abducens nucleus.

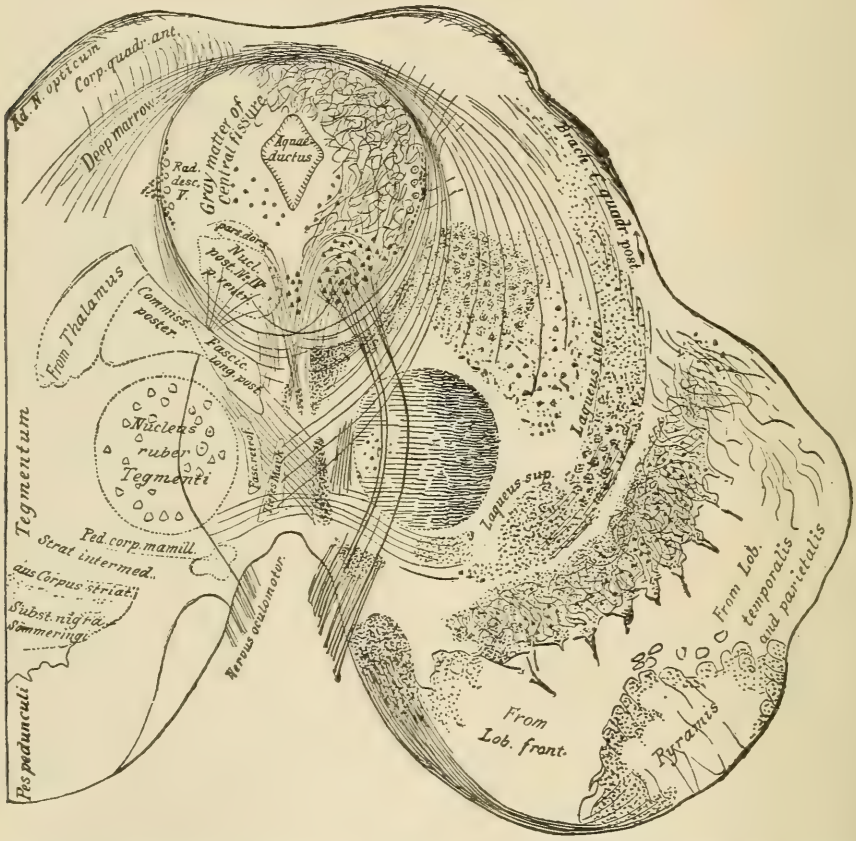


FIG. 120.—Schematic cut behind the anterior quadrigeminal bodies through the nucleus of the oculomotor. Instead of nucleus *Nr. IV* read *Nr. III*. (*After Edinger.*)

The fibres pass dorsally around the nucleus of the abducens nerve, then ventrally and appear at the posterior edge of the pons, at the side of the olive.

The *intermediate nerve* (N. *intermed. Wrisbergii*), classified as a minor part of the facial nerve because so closely associated with it, has partly sensory functions (fibres of taste), partly secretory (through the chorda tympani). Its sensory fibres from the geniculate ganglion end probably in the sensory end-organ of the glosso-pharyngeal nerve.

The *auditory nerve* (*nervus acusticus*, N. VIII), cf. auditory paths. (See Fig. 122.)

The *glosso-pharyngeal nerve* (N. IX), a mixed nerve. The motor fibres arise in two nuclei:

1. The *dorsal* nucleus in the floor of the fourth ventricle, and
2. The *ventral* nucleus (or *ambiguus*), dorsal from the olive.

The *sensory* fibres arise in both ganglia of the nerve (G. superius and petrosum) and pass as sensory roots into the medulla oblongata beneath the facial and auditory nerves behind the olive.

Here the fibres divide into ascending and descending branches. The ascending end in the *nucleus alæ cinereæ*, the descending (the so-called *tractus solitarius*) in the adjoining nucleus tractus solitarii.

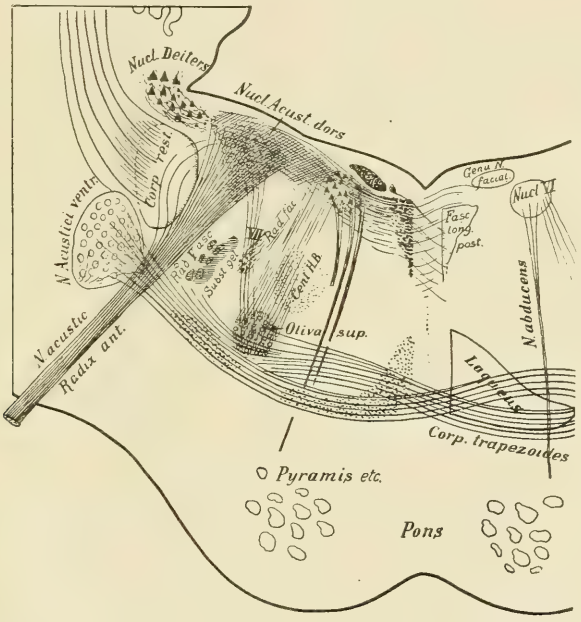


FIG. 121.—Cross-section in the region of the origin of the abducens. (*After Edinger.*)

The *pneumogastric nerve* (*Nervus vagus*, N. X), a mixed nerve. The *motor* fibres arise in the same nuclei as those of the *glosso-pharyngeal* nerve, that is in the *nucleus dorsalis* and in the nucleus *ambiguus*. The sensory fibres arise in both ganglia of the vagus nerve (G. jugulare and nodosum), pass below the ninth nerve, back of the olive into the medulla, and end in the same sensory end nuclei (nucleus alæ cinereæ and nucleus tractus solitarii) as those of the glosso-pharyngeal nerve.

The *spinal accessory nerve* (*nervus accessorius*, N. XI), a pure motor nerve. It rises from a collection of cells that begins in the caudal extension of the nucleus ambiguus (cerebral part of the accessory nucleus) and can be traced to the seventh cervical segment (spinal part of the accessory nucleus).

The latter is situated at the base of the lateral horns, and in the dorsolateral part of the anterior horns of the spinal cord.

From this nucleus arise 9–13 root-fibres, which leave the medulla oblongata, resp. the cervical white matter (Fig. 124).

The *hypoglossal nerve* (nervus hypoglossus, N. XII), a pure motor nerve. The nucleus lies in the floor of the fourth ventricle, next to the raphe. The fibres pass ventrally and leave the medulla in 10–15 root-fibres, between the olive and pyramid (Fig. 125).

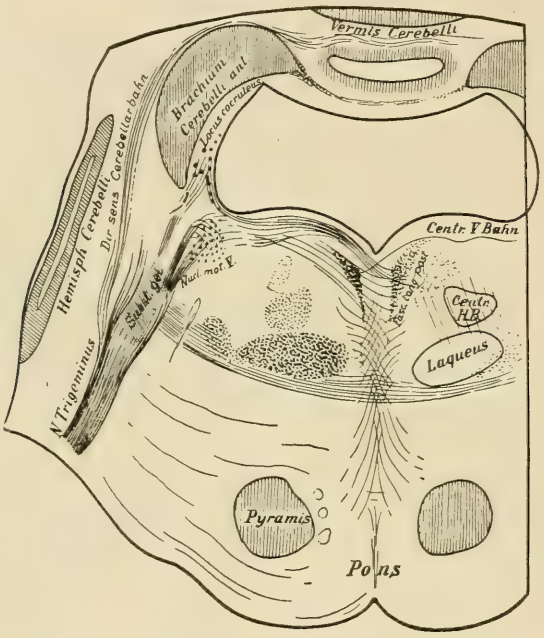


FIG. 122.—Motor nucleus and root of the trigeminal. (*After Edinger.*)

6. General Relation between Structure and Function of the Brain

(a) THEORY OF PROJECTION; PATHS AND FIELDS OF PROJECTION

The cortex receives stimuli from the sensitive parts of the surface of the body (the entire skin, joints, muscles, so-called higher sensory surfaces, as retina, organ of Corti, etc.), and sends out stimuli into parts that may be moved volitionally, to the muscles, and to certain glands.

The paths which connect the cortex with the periphery are called *projection* paths.

There is no *uninterrupted* connection between the cortex and a sensory or muscular end-organ. *Between the cortex and periphery gray masses* (e. g., of the spinal cord) *are interposed.* In the cells of these interposed gray masses the sensory nerves end and the motor arise.

The cells of these gray masses are connected with the cortex through ascend-

ing paths. There are then in the projection paths, between periphery and cortex, *one or more gray-matter way-stations*. So all *sensory* nerves of the *trunk* and the *limbs* starting from the periphery have their first end in the gray matter of the spinal cord or of the medulla oblongata (sensory end-nuclei). And all *motor* nerves for trunk and limbs have their *origin* in the gray matter of the spinal cord (anterior horn cells, motor origin cells).

But the *cerebral* nerves are wholly analogous to these. They, too, have their sensory end-nuclei and their motor origin nuclei in the gray matter lying next to the fourth ventricle and the aqueduct of Sylvius. These nuclei, too, are aggregations of ganglia cells, which—in *centripetal* directions—receive the impulses of the peripheral sensory neurons and transmit them to their axons leading to the cortex, or—in *centrifugal* directions—their axons, themselves, become motor nerves, passing to the periphery.

Just because this gray matter of the medulla and the quadrigeminal region receives sensory roots in the end-nuclei, and sends out motor roots from origin nuclei, one has named this cerebral region in consequence of its far-reaching analogy with the function of the spinal cord, the *spinal* cerebral region. This gray matter in the floor of the fourth ventricle and aqueduct, we can, when taken together with the nuclei of the posterior column of the medulla and the gray matter of the spinal cord (anterior and posterior horns), look upon as the *first* station of combined sensory and motor processes, and as *primary* centers of these functions. These primary centers represent the *first projection* of the periphery in the central nervous system.

Any lesion of these primary centers interrupts the functional capacity of some peripheral part, whether it be sensory or contractile. About the term projection, see below.

For the further connection of these primary centers with the gray matter of the cortex, we must consider separately, the motor and the sensory system.

For the main paths of motility (direct motor paths), there is no further way-station between muscles and cortex, besides the subcortical gray matter we have just mentioned, which contains the origin nuclei of the motor nerves in the midbrain, medulla oblongata and spinal cord. For more detailed information about these origin nuclei cf. page 458.

From the cortex to these nuclei there is an uninterrupted conduction path, *the central motor path.*

Its *region of origin in the cortex* represents, for the second time, all voluntary motor processes (projection of the *second* class, cortical center), in a circuit still to be described. For the voluntary movements, we have, then, 1. a cortical projection, 2. a subcortical projection (the latter for the cerebral nerves in the gray matter of the quadrigeminal bodies, the pons, medulla oblongata, and for the nerves of the trunk and limbs in the gray matter of the anterior horn of the spinal cord). Both projection centers are connected by motor projection paths. The main path for the voluntary move-

ments of the limbs and the trunk is also called the *pyramidal tract* (cortico-spinal path). It rises in the upper two-thirds of the anterior central convolution, and its immediate vicinity, and in the paracentral lobule and passes, without interruption, through the centrum semiovale, the inner capsule (anterior two-thirds of the posterior limb), through the foot of the cerebral peduncle and the pons into the medulla, at the lower end of which its greater part undergoes decussation. The part that has decussated passes into the lateral tracts of the spinal cord, and ends in the anterior horn on the same side, from the cells of which the anterior roots, the motor nerve rises. The non-decussating fibres pass away in the ventral bundle,

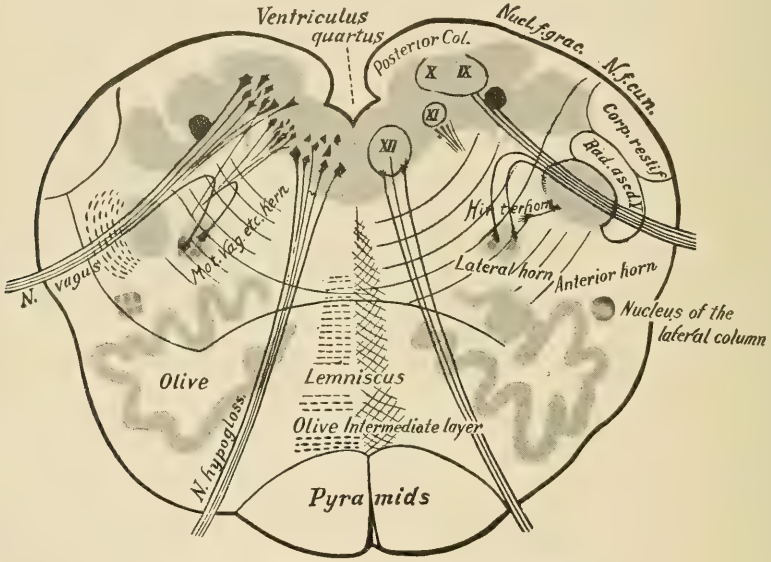


FIG. 123.—Exit of the vagus. (*After Edinger.*)

and reach the anterior horn on the same side. The pyramidal tract is the longest of all conducting paths.

The corresponding motor path for the *cerebral* nerves, the *cortico-bulbar* path, arises in the lower third of the anterior central convolution, passes through the white matter, through the genu of the inner capsule, the foot of the peduncles, and ends, with much decussation, in the contralateral nuclei of the cerebral nerves.

Besides these important *direct* motor paths there are *indirect* paths, interrupted in the *red nucleus*, and very important in the higher animals, but less so in man. There is no unanimity of opinion as yet as to whether the central path, that leads to the red nucleus, comes straight from the cortex, or is interrupted in the thalamus, or whether it first makes a large detour about the cerebellum, as do the pyramidal tracts which pass within the tract of the crusta through the internal capsule and the cerebral peduncle to

the pontile nuclei (decussated) and into the cerebellum and through the superior peduncles of the cerebellum to the red nucleus.

Things are more complicated for the *centripetal* paths, as well for general, as for special sensation. Between the first endings in the gray matter of the end-nuclei, and their final endings in the cortex, *if we ignore further interruptions*, one way-station, at least, is always interposed; this is formed by the gray nucleus of the optic thalamus with its appendages (both geniculate bodies) and the gray matter of the quadrigeminal bodies.

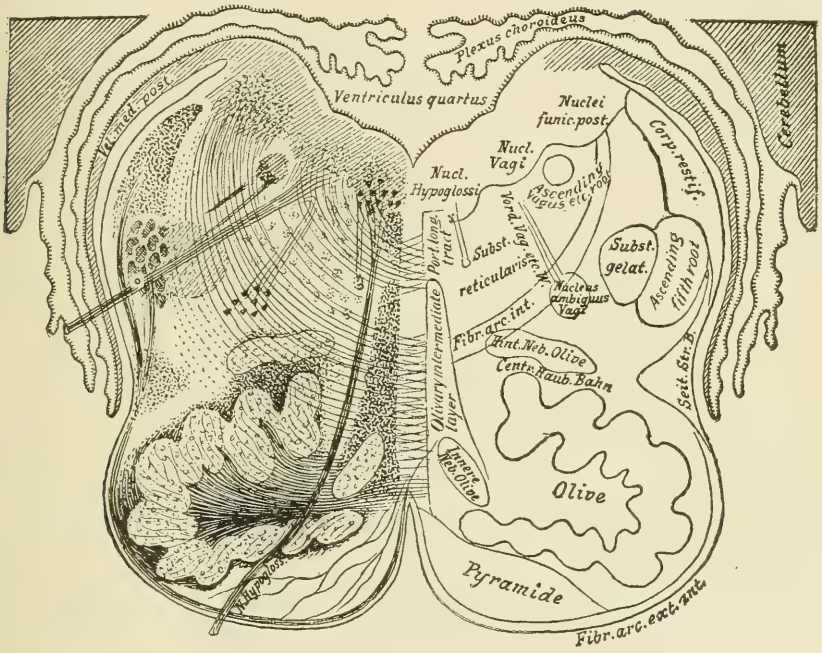


FIG. 124.—Nucleus and root of the hypoglossal nerve, nucleus ambiguus and root of the vagus. (*After Edinger.*)

From the sensory end nuclei arises a second centripetal path, which, for the most part, passes into the *medial* and *lateral fillet* and ends in the optic thalamus with its appendages and in the quadrigeminal bodies. Therefore the thalamus and the quadrigeminal bodies represent a second sensory projection. From the thalamus and the quadrigeminal bodies arises, then, a third path which ends in the cortex of the cerebrum, so that the cortical sensory projection is at least the third projection. (Cf. also page 491.)

Although, according to its *embryological* development, the ganglia cells of the retina do not correspond to the spinal ganglia, nor the optic nerve, to a posterior root, but rather represents the connection of two cerebral parts (the retina is a cerebral part that has been pressed forward), *physiologically* the same observation that we have just made with reference to the other

centripetal nerves may be applied to the connection between retina and cortex. Even the fibres of the optic nerve, which arise in the cells of the retina, find their first ending in a subcortical nucleus, its primary center, only this lies in the interbrain or midbrain (the *second* station of the rest of the sensory paths), namely, in the external geniculate bodies of the optic thalamus and the anterior quadrigeminal bodies. Hence the central path goes through the optic radiation into the cortex of the occipital lobe (cf. later visual pathways). Here also there is a primary subcortical and a secondary cortical projection.

FIG. 125.—Nucleus of the *N.* hypoglossus (Weigert stain).

The *olfactory* path demands separate treatment. The axons of the peripheral nerves end in the olfactory bulb; from here a second path goes, especially to the gray matter of the olfactory trigone and the anterior perforated substance; from here a third path, which ends in the cortex of the gyrus hippocampi and in the hippocampus major. Therefore for the optic and olfactory nerves, between which and the other sensory nerves, in respect to their embryological development, there is not the slightest analogy, the

same law holds good, viz.—that between the periphery and the cortical center gray matter intermediate stations are interposed.

Therefore we can say: *All the sensitive surfaces of the body and all its organs that produce motion, are projected upon the cortex by the help of one or more intermediate stations of gray matter.*

The word projection is not to be understood literally, even in its application to the first projection in the spinal cord, and certainly not when applied to cortical projections—not in the sense, that for every point in the periphery, there exists a co-ordinate point in the cortex, and that the arrangement of the separate cortical elements is a replica of that of the peripheral sensory and muscular elements. Such correspondence between periphery and cortex does not exist. For instance, not each separate muscle has its own center in the cortex, but certain groups of muscles for definite related movements are represented there. [In other words there is a center for movements rather than for muscles.] But *in broad outline*, the projection does exist, in so far as various sense-organs, and various sensory and motor processes of the limbs are co-ordinated with various regions of the cortex. And even if, within such a cortical region, corresponding to the motor processes, say, of an upper extremity, the arrangement does not repeat the muscular scheme in absolute detail, on the other hand, the entire region is not as a mass co-ordinated to the entire limb, but parts of it correspond to certain groups of muscles. In this broader sense, the conception of a projection of the body, on the cortex, is justified.

Those cortical regions, that receive projection paths, or from which they emerge, are called fields of *projection.* Their destruction is marked, therefore, by the destruction of the motor processes or sensory processes of definite perceptive or effector organs; their injury without destruction by convulsions or other phenomena of motor irritation, or pains or other sensory phenomena of irritation (elementary hallucinations, for instance).

(b) The Mnemic Cortical Fields and the Theory of Association Fields

The reception of centripetal stimuli in the projection fields, *when this innervation has reached a certain level*,[1] is the cause of *perception.* The excitation of motor fields of projection, at a certain level leads to *innervation* and subsequent *movement.*

The cerebrum has another broader function—that of retaining lasting traces of sensory or motor innervations that once existed (residues or remnants, the possession of the memory). These remnants not only make

[1] Not every stimulation of the cerebrum is connected with consciousness, though it is to be assumed, that, in man, no consciousness is possible without corresponding stimulation of the cerebrum.

possible *conscious* recollection of perceptions and movements, but also, even without leading to *conscious* reproduction, as *purely material* possession make it possible for the nervous system, to perform repeated operations with greater ease, certainty and perfection; upon them therefore depend *dexterity*.

This is the *mnemic* function of the cerebrum. On the other hand, it produces associations among these residues; this is the *associative* function of the cerebrum. The smell of a rose, for instance, rouses in consequence of such association, a complete image of a rose.

It has been long debated whether the mnemic-associative functions are bound to *the same* cortical regions as perception and innervation, or if they have their own peculiar cortical fields.

According to a widely held opinion all parts of the cortex receive projection paths, or send them forth, so that the entire cortex is divided into projection fields. The elementary mnemic possession belonging to every projection field, that is, the remnants in any one sense field, would, eventually be localized within the same cortical region, but may be in other layers as perception and innervation, so that, for instance, the association among elements of the same sense fields, say among the optic elements, would take place in the same cortical regions, the association, on the other hand, among the remnants of various sense fields (optic, tactile, etc.), would be brought about by the long associative fibres. Then, the same cortical region that receives the optic radiation would possess *optic memories*. In this view, it is not necessary to assume that the same *nervous elements* serve for perception, innervation, and memory, as this is, indeed, very improbable; the remnants might be registered in the *same large cortical region*, and yet be bound to different *cell-fibre complexes*.

Opposed to this is *Flechsig's* theory, which accepts only a small part of the cortex as projection fields. According to him, only these are in communication with the periphery through in- and outgoing paths. *The rest of the cortex has no such connection with the periphery*. It is occupied by the *association fields*, that have connections only with the projection fields (associative fibres) and with the opposite hemisphere (commissural fibres) and carry on the associative and higher mnemic functions. Furthermore Flechsig divided the entire cortex, according to the time when myelin develops into 36 different fields, of which those first maturing (primordial regions), 12 in number, constitute the *projection fields*, the majority of those maturing later (intermediary regions), 16 in number, and the 7 which mature last (terminal regions) exclusively constitute association fields (Fig. 126).

Flechsig's projection fields include as most important, both central convolutions, beside the paracentral lobe, the adjacent part of the first frontal convolution and the gyrus fornicatus, the transverse convolution of the first temporal convolution, with a small part of the latter, the vicinity of the calcarine, also little fields of the surface of the occipital lobe, the uncus of the

hippocampal gyrus, and the inner surface of the temporal poles. All the rest of the surface of the brain, and especially, *both inferior frontal convolu-*

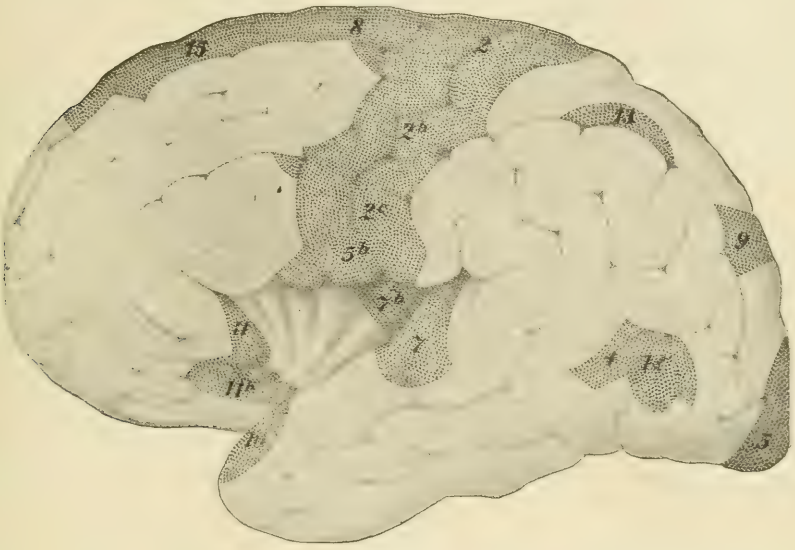


FIG. 126.

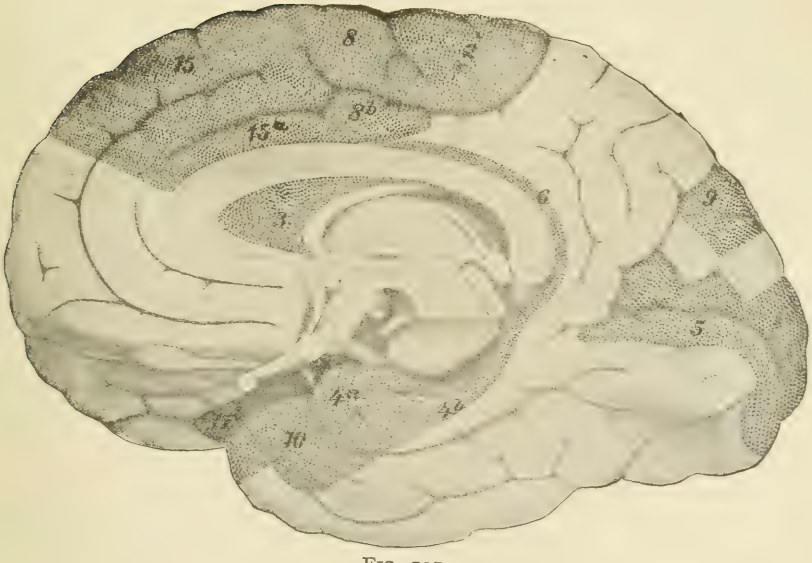


FIG. 127.

FIGS. 126 and 127.—Projection and association fields. According to Flechsig corona radiata fibres are certainly proved to exist only in the dotted regions, not in the regions not dotted. The remainder of the first temporal convolution and of the Gyr. hippocampi are still under consideration. The other undotted regions are, according to Flechsig, association centers. (*After Flechsig.*)

tions, the insula, the rest of *the temporal and occipital lobes and the* entire parietal lobe, are, according to Flechsig, association fields (Fig. 127).

In Flechsig's theory two theses are included that demand separate treatment: first, the purely *anatomical*, that only part of the cortex receives and sends out corona radiata fibres, that only this part is connected directly with the periphery and that the remaining portions of the cortex, are *not*, therefore, projection fields.

The second thesis is *physiological*, relating to the *function* of the fields in which corona radiata fibres are absent. These fields serve for the *association* between optic, tactile, etc., processes, and, at the same time, for the higher, even the highest psychic processes.

So far as the *anatomical* question is concerned, it must be granted Flechsig, that the theory of equal distribution of projection fibres on the cortex can *not* be maintained. *The majority of the most important motor and sensory tracts actually crowd together into narrow regions, lying in and about Flechsig's projection fields.* Even if the other regions are not wholly free of projection fibres, and this or that boundary of Flechsig is, as yet, disputable, we must, at any rate, recognize the important separation of these regions which are very rich and those which are very poor in corona radiata fibres, whereby the fundamental division of Flechsig must be conceded, at least a relative justification.

A second question is, whether these regions in which corona radiata fibres are scanty, are really seats of *the highest associations*. Another possibility is, that they have elementary mnemic functions, and as *Exner*, *Nothnagel* and *Ziehen* thought, that they are to be regarded as *memory fields*.

Then, in the neighborhood of every projection field, the corresponding remnants of any one kind of sense impressions would have their substratum, i. e., in the neighborhood of the acoustic projection field, the acoustic remnants, in the neighborhood of the optic projection field (medial side of the occipital lobe), the optic remnants (convexity of the occipital lobe). The regions poor in corona radiata fibres, would then be mostly *memory fields*, nevertheless the intra-sensory associations would take place within them, which, as all authors agree, take place within the cortex. *But associations among the various sensory and motor areas, and all higher associative complexes*, would, as Wernicke supposed, be carried on through the large association paths running in the white matter.

For this latter supposition much experimental evidence may be adduced.

In the entire question, which can by no means be, as yet, satisfactorily decided, the following point of view seems to be indicated as a compromise:

Certain cortical regions richly supplied with corona radiata fibres are connected with receptor and effector organs: the *projection fields*.

There are further cortical areas poorly supplied with corona radiata fibres, without important connection with the periphery.

But the distribution of perception and innervation, of memory and association is not such, that the fields of the former serve *only* for projection,

and *only* those of the latter for memory, and the latter, moreover, for the higher associations but the fields *of the former kind*, serve for retaining the remnants *as well* as for projection.

The fields of the latter kind serve almost exclusively for remnants, hence deserve the name, mnemic fields. The association between the remnants of *the same* sense takes place all over the cortex.

The associations between remnants of various senses, and all higher complexes are made possible only by the long association fibres. This higher association is bound to the systems, which by association fibres, represent projection and memory fields combined.

Hence, no part of the cortex is wholly free from remnants. Granted only, that certain parts serve *as well* for projection, while others do not, we may distinguish the latter as *purely* mnemic fields, from the projection fields. And the higher associative processes are not carried on in separate cortical fields but demand the long association fibres, which unite various cortical fields in a common activity.

(c) Association Paths

The *short* association fibres which run within the cortex (intracortical) or unite neighboring gyri (arcuate fibres), vary greatly in arrangement and direction in different parts of the cortex.

The *long* association fibres, which unite distant parts of the cortex, run, the longer they are, the deeper into the white substance. The knowledge we have of them is still very limited.

Certain conspicuous aggregations of long association fibres in compact bundles have received individual names. But one must not forget that the many, more scattered fibres with association functions, are not included therein.

These bundles consist not only of long fibres, which run from beginning to end in the bundle, but, in their course, many fibres from the cortex enter, and many pass out (Fig. 128).

Only the *most important* pathways of the long association fibres will be mentioned and named:

1. *Uncinate fasciculus.* Connects the orbital surface of the frontal brain with the pole and anterior portion of the temporal lobe.

2. *Superior longitudinal fasciculus* (or arcuate). From the frontal brain to the parietal-occipital lobes and the *posterior portion of the temporal lobe.*

3. *Inferior longitudinal fasciculus*, ending in the vicinity of the posterior horns, as the outermost of the three sagittal medullary layers, that surround the posterior horn (cf. Figs. 143 and 144). It was considered formerly a mere bundle of association fibres between temporal and occipital lobes, but is a mixed pathway. It takes also many projection fibres to the

visual sphere, contains optic radiations and even corpus callosum fibres, besides association fibres.

4. *Occipito-frontal fasciculus*, adjacent to the caudate nucleus, a mass of fibres also passing from front to back.

5. *Cingulum.* Runs in the white matter of the gyrus fornicatus, beginning in the anterior part of the frontal brain, ending in the occipital lobe, consisting of sagittal fibres, which, mostly, connect adjacent cortical areas, but contain single fibres running through its entire length.

6. The external and extreme capsules also contain sagittal association fibres. See still other bundles of association fibres in Monakow's scheme.

(d) The Commissures, especially the Corpus Callosum

The commissural fibres connect cortical parts of both hemispheres.

1. The most important commissure is the corpus callosum. The mass of fibres included in the center is known as the body.

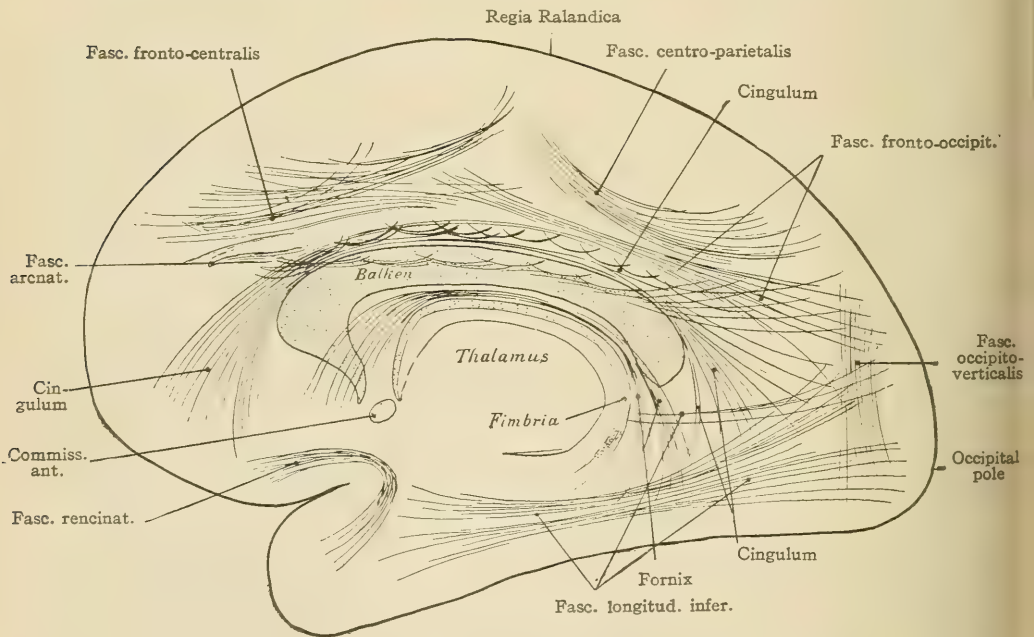


Fig. 128.—Scheme of the most important association paths. (*After v. Monakow.*)

The anterior curved portion is called the genu (knee); the posterior thickening, the pad or splenium; the middle part the body.

It connects, by means of intra-hemispherical corpus callosum fibres, symmetrical and asymmetrical cortical areas. This fibrillation is mixed with the projection and association fibres, and constitutes a great part of the white matter (Fig. 129).

The knee (genu) sends its fibres into the frontal brain, as the *anterior*

forceps; the splenium into the occipital and temporal lobes as the *posterior forceps*, the fibres of which, for the most part, pass over into the *tapetum* of the posterior and inferior horns of the lateral ventricle. The body of the corpus callosum connects the central parts of the hemispheres, including both central convolutions.

If one considers, that in the centrum semiovale projection and commissural fibres run side by side (see scheme), but separate at the lateral edge of the lateral horn, that then the projection fibres pass into the inner capsule, whereas the commissural fibres, forming the corpus callosum, pass over the ventricle, to the other side, it is clear, that in focal injuries there is a distinction, *but lately discovered:* a focal injury (*I*, Fig. 129) in the centrum semiovale

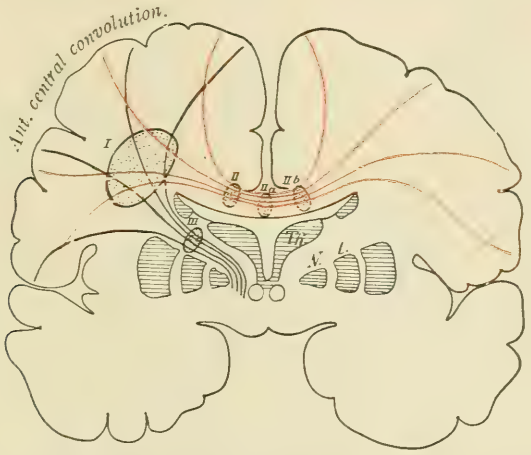


FIG. 129.—Scheme of the fibres in the corpus callosum (red). In the frontal plane only connections of symmetrical regions could be indicated. The projection fibres black. *I*, *II*, *IIa* and *b*, *III*, foci. *I* in the white matter affects fibres in the corpus callosum and projection fibres. *II*, *IIa* and *IIb* only fibres in the corpus callosum. *III* in the inner capsule projection fibres only. *Th*, thalamus; *N.l.*, lenticular nucleus; anterior central convolution.

strikes projection fibres and fibres of the corpus callosum, a focal injury in the inner capsule (*III*) *only* projection fibres, a focal injury in the corpus callosum (*II*, *IIa*, *IIb*), *only* commissural fibres. Since (see under apraxia) the left hemisphere, through the corpus callosum, affects the innervations of the right hemisphere, a focal injury (*I*), will cause besides paralysis of the right extremities, dyspraxia of the left. A focal injury in the inner capsule, on the other hand, produces only paralysis of the right, in the corpus callosum only dyspraxia of the left extremities. Extensive interruption of the fibres of the corpus callosum, probably especially of the central third, produces a local symptom: dyspraxia of the left upper extremity—it does not matter whether the interruption is in the left hemisphere, the corpus callosum itself, or in the right hemisphere. By the great crowding of fibres in the corpus, a lesion therein will, of course, be most fateful for praxia.

Of great importance in the production of motor aphasia, is the interrup-

tion of the anterior fibres in the corpus, since the frontal speech center influences the right brain centers of the 7th and 12th nerves through the corpus fibres.

So, too, interruption in the splenium fibres either in it or within the forceps, and the white matter of the temporal and occipital lobes can help to cause alexia and mind blindness (see sections dealing with these subjects).

2. The anterior cerebral commissure connects the basal parts of the temporal and frontal lobes.

3. The lyre of fornix (Lyra Davidis) connects the hippocampal horns.

7. Clinical Localization in the Left Hemisphere

Fortunately pathology, *roughly speaking*, is not seriously affected by differences in the views discussed in pages 467–472.

For, even if the answer is left open, whether the association between various qualities (optic, tactile, acoustic, etc.), is secured by means of specialized cortical fields, orby long *associative pathways*, on one point the adherents of both views agree, that lesions of certain areas in the cerebrum, if cortex *and white matter* be considered *together*, produce no massive, striking symptoms of lost functions in the sensory and motor realms, but cause serious mnemic disturbances and loss of associative processes, that, on the other hand, lesions in other areas are first chiefly characterized by paralysis or anæsthesia. Thus, destruction of the anterior central convolution causes hemiplegia, of the calcarine region, hemianopia; on the other hand, a lesion in the third frontal convolution, the posterior third of the first temporal convolution, of the parietal lobe and its transition into the occipital lobe (in so far as there is no injury of the *pathways* leading to the primary projection centers), causes no paralysis or anæsthesia, but disturbances in speech, writing, reading, agnosia, apraxia, respectively (cf. page 518, ff). Whereas Flechsig and his school connect these mnemic-associative disturbances mainly with cortical lesions, we trace them mainly to injury of the associative paths that pass below the cortex.

So we can say with some degree of certainty what clinical symptoms of lost function will follow upon the destruction of *entire* parts of the cerebrum, including cortex and white matter, i. e., of entire lobes, or parts of lobes, though the question as to how far the *cortical* elements themselves, or the *association fibres* passing through the white matter, are implicated, is, as yet, in many respects, debatable. If one combines such a purely clinico-pathological localization with the known results of electrical stimuli used to experiment upon motor areas, the following localization appears.[1]

For the more detailed localization of motor functions, the evidence

[1] Only those symptoms have been described, which may be regarded as symptoms resulting directly from injuries, not conditioned by the effects of vicinity or distance. The entire survey, because of its brevity, can naturally give only a *summary* orientation.

obtained by means of electrical stimulation, is of the greatest importance. It has been shown that, in man also, the easily stimulated points are situated, in overwhelming majority in the *anterior central convolution* (Ca), that the *lower third* belongs to the muscles of the head, the middle third to those of the upper extremity, the upper third, together with the paracentral lobe, to those of the lower extremity.

The posterior two-thirds of the inferior frontal convolution (possibly

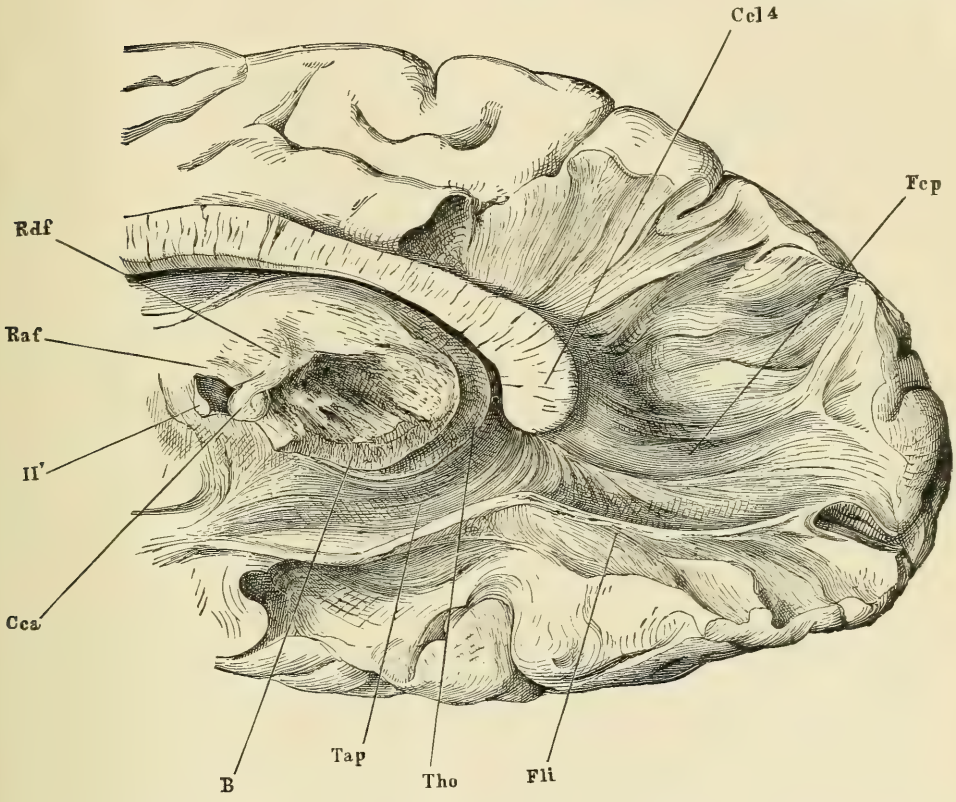


FIG. 130.—The corpus callosum and its radiations. By breaking off with a forceps, the; radiation of the posterior end of the corpus callosum is indicated. *Ccl*4, Splenium; *Tho*, thalamus; *Tap*, tapetum; *C*, *ca* corpus candicans; *B*, place where the cerebral peduncle is cut; *Fcp*, forceps) *II'*, optic nerve; *Raf*, fornix; *Rdf*, Vicq d'Azyr stripe; *Fli*, fasciculus longit. inf. (*After Edinger.*

also the lower third of the middle frontal convolution) besides the adjacent part of the lower fourth of the central convolution, together with the anterior part of the island of Reil form the *frontal speech region*, the destruction of which causes *motor aphasia.*

Focal injuries at the *base* of the frontal brain cause, if they destroy the olfactory bulb and tract, *inability to smell* on the same side.

Destruction of the lower fourth of the *anterior* central convolution, alone, causes paresis of the contralateral muscles of the tongue, palate, lips, cheeks

and of those used in mastication, which, however, as all these muscles are innervated from both hemispheres, is not very serious, and (except for the paralysis of the contralateral genioglossal muscle) is not permanent. Consequently, only a *moderate* degree of dysarthria remains.

Serious paralysis of the muscles mentioned, *on both sides*, as well as of those of the throat, hence, also anarthria or *lasting serious dysarthria* only appear, when, at the same time, the corresponding area of the right hemisphere is destroyed (cortical pseudo-bulbar paralysis).

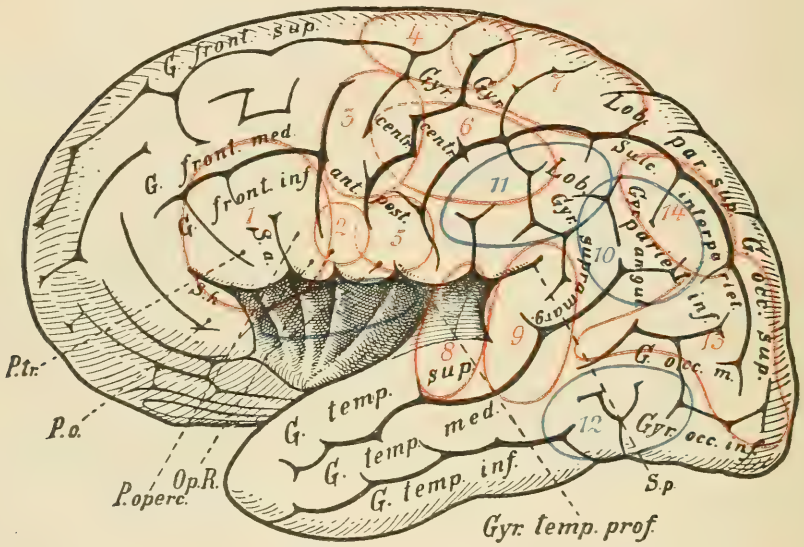


FIG. 131.—Sketch to explain the effect of variously situated foci in the left hemisphere. The blue lines indicate that subcortical foci corresponding to the demarcated region cause the loss of function concerned. 1, Motor aphasia (because of the illustration showing the third frontal convolution in direct upward view and not shortened, the frontal speech region appears unproportionately large); 2, paresis of the lingual, facial, masseter, deglutition and laryngeal muscles on the opposite side (transitory except for the tongue muscles); 3, paralysis of the arm and the hand; 4, paralysis of the leg; 5, 6, 7, disturbances of sensibility in the face, arm, leg (in 6 also paralysis of touch); 8, if destroyed bilaterally, deafness—if on the left side, pure word deafness (?); 9, sensory aphasia; 10, near to the convexity, alexia and agraphia—if deeper near the medial region, pure alexia; 11, near it, amnestic aphasia—deeper, apraxia; 12, amnestic aphasia; 13, if bilateral, mental blindness (visual agnosia) which, however, may occur also because of various other combinations of foci, besides amnestic (especially optic) aphasia; 14, déviation conjuguée; 15, insular aphasia (blue). *P.tr.*, pars triangulars of the third frontal convolution; *P.o.*, pars orbitalis of the third frontal convolution; *P.operc.*, pars opercularis of the third frontal convolution; *Op.R.*, operculum Rolandi; *S.h.*, *S.a.*, *S.p.*, ramus horizontalis, ascendens, posterior fossæ sylvii.

Lesions in the first and second frontal convolution may disturb, some say, equilibrium in walking, as do lesions in the cerebellum (frontal brain ataxia).

Regarding a center for eye movement, to be found in the frontal brain, see page 478.

The central two-fourths of the anterior central convolution contain, first of all, the motor center for the upper extremity on the other side. From

above downwards occur in natural sequence the centers for shoulder, arm, hand, finger.

The upper fourth of the anterior central convolution, and the anterior part of the *paracentral lobule* lying on the median surface contain the motor center for the legs. The posterior central convolution, and a part of the adjacent superior parietal lobe, is the seat of *sensibility* of the opposite half of the body (arranged from above down, like those of motility) and the posterior central convolution is preeminently the seat of *perceptions of movement and position, of the sense of place and space* and the corresponding concepts. These perceptions and representations reach over also into the *principally motor* area of the anterior central convolution (Fig. 131).

In contradistinction to this, the area for touch and pain sensations extends over the posterior central convolution, backwards, at least in the front parts of the superior parietal lobe. Destruction of the middle third of the posterior central convolution and the directly adjacent part of the parietal lobe produces *paralysis of touch.*

The middle third of the superior temporal convolution, together with the *transverse convolution of the temporal lobe* (gyrus transversus or profundus) represent the auditory center, the destruction of which, because of the demi-decussation of the acoustic fibres causes deafness, only when it is *bilateral.*

Destruction of the third of the superior temporal convolution, lying behind it, and of the anterior adjacent parts of the supramarginal convolution causes *sensory aphasia.* Lesions in the angular gyrus cause *disturbances in reading and writing* with *difficulty in finding words.*

Lesions in the *depths* of the angular gyrus near the median and basal surface of the brain cause merely disturbances in reading and difficulty in finding words.

Large lesions in the *white matter* of the supramarginal gyrus, of the angular gyrus, and possibly also in the white matter of the upper parietal lobe cause, in addition to these, apraxic disturbances. See page 544.

Lesions at the base of the posterior half of the temporal lobe and the adjacent parts of the occipital lobe cause particularly great difficulty in finding words (amnestic aphasia) (Fig. 132).

Focal injuries in the cortex and white matter of the *convexity* of the occipital lobe affect visual memories, the appreciation of forms and spatial relations and the assimilation of visual sensations. In order to cause mind blindness, a *left*-sided focal injury will suffice in some people; in the majority, however, a bilateral occipital focal injury is necessary to cause serious mind blindness.

Focal injuries on the *median surface* of the occipital lobe, which destroy the vicinity of the calcarine fissure, cuneus, lingual gyrus, and, at the base the fusiform gyrus, cause *half-blindness.*

Focal injuries, which in the white matter of the occipital lobe or even of

the parietal lobe, include the optic radiation, cause half- or quarter-blindness.

In the *angular gyrus* lies a projection center for the movements of both eyes in the opposite direction. *Irritation* there causes a deviation of the eyes towards the other side (the patient looks away from the side with the focal injury), and frequently, simultaneous turning of the head.

Focal injuries, however, in the pons, between the oculomotor and abducens nuclei, cause *paralysis of the "looking process"* (Blicklaehmung), not to be confused with deviation; the eyes can not be led across the middle line, nor towards the side of the focal injury.

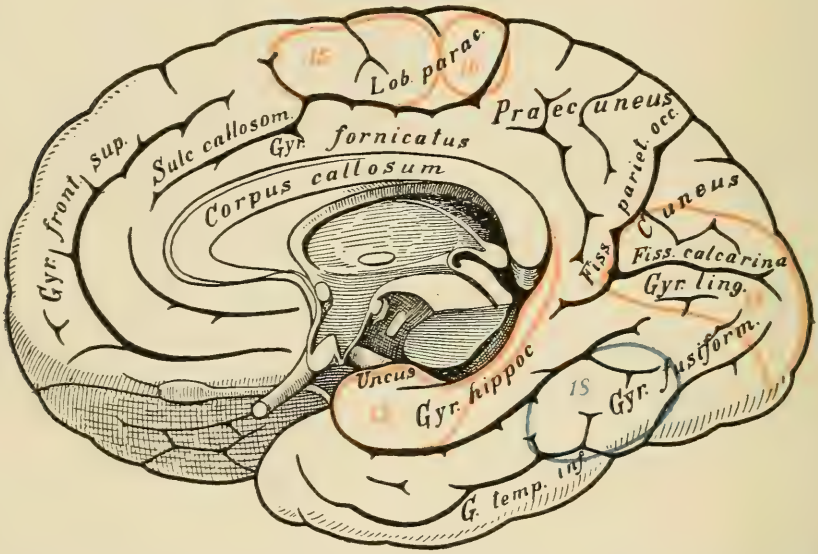


FIG. 132.—Foci on the median surface of the left hemisphere. 15 (red), Paralysis of the leg; 16, disturbance of sensibility in the leg; 17, hemianopia towards the right side; 18 (in the white matter) amnestic aphasia; 19, is said to produce disturbances in smelling.

Paralysis of the center of eye movement in the angular gyrus leads possibly, since from here simultaneously the inhibition of the antagonists takes place, to preponderance of contralateral looks (the patient looks towards the focally injured side). A second center of the eye muscles in the frontal brain, which has been proved to exist in higher animals, is assumed by some authors to exist in man, also, at the foot of the first and second frontal convolution.

The hippocampal gyrus, uncus and hippocampus major are brought into relation with the senses of smell and taste, with the latter, especially the back part of the fornicatus gyrus.

Of the functions of the *lenticular nucleus* and *caudate nucleus* nothing certain is known; injury of the *optic thalamus* with its appendages, as intermediate station for all sensory stimulations, even for the higher perceptions,

causes serious disturbances in sensibility. Destruction of the *outer geniculate body*, hemianopia. Destruction of one of the inner geniculate bodies leads only to serious *auditory* disturbances, when the auditory path on the opposite side is injured somewhere in its course.

8. Differences in Function of the Two Hemispheres

While for projection functions—movement, sensation, seeing, hearing, tasting, smelling—the right hemisphere is naturally equal to the left in all men, it takes a far smaller part in the higher mnemic and associative functions. This holds good, at least, for right-handed people, ca. 90% of the total. The opposite is true of the 4 to 5%, of left-handed people. Both hemispheres are of equal importance in the ambidextrous, 5 to 6%.

The right hemisphere in the right-handed is so far from indispensable for functions of speech, that destruction of the right frontal brain, the right temporal lobe, the right angular gyrus, very seldom causes disturbances in speech, writing or reading, so that these functions can be fully performed without the right hemisphere.

On the other hand, intactness of the right hemisphere does not protect persons who have suffered extensive lesions in the left speech area, from lingering speech disturbances, that are often serious and permanent. In the course of time, however, the right temporal lobe may vicariously represent and take the place of the left, and, to a certain extent, obliterate the aphasic disturbances.

Likewise the left hemisphere is more important for *praxia;* through lesions in certain parts of the left hemisphere, not only the right, but also, to a certain extent, the left upper extremity, loses the power to perform movements from memory, to imitate movements, etc. For the simpler manipulation of objects, the memory movements of the right hemisphere suffice, as a rule. Even for memories of form and color, precedence must be assigned to the left hemisphere, and especially for higher association of all simpler memories, and the *spontaneous* awakening of memories.

While lesions of the central convolutions and of the auditory, visual, olfactory and gustatory areas in the right hemisphere cause identically the same clinical symptoms of lost function on the left side, as those of the corresponding parts of the left hemisphere cause on the right side, the right frontal lobe and large parts of the right temporo-parietal lobe are marked as "*silent*" parts of the brain (lesions of the occipital lobe are noticeable through the interruption in the optic radiation usually resulting), by which is meant, that mnemic-associative disturbances that are caused by focal injuries in the corresponding areas on the left side, are only slightly noticeable when the lesion is on the right side. Naturally with refinement of methods, even the silent parts of the brain will begin to speak, and it must

be assumed, that they, only in a far less degree, have a mechanism similar to the corresponding parts of the left hemisphere. When *added* to left-sided lesions, the corresponding right-sided lesions are clearly brought into evidence even to-day.

9. Cerebellum

The *cerebellum* demands special attention. Cf. Figs. 114 and 115.

Of the morphologically important relations we emphasize only that the cerebellum is connected with the rest of the central nervous system through three pairs of crural connections:

1. Crura cerebelli ad cerebrum or ad corpora quadrigemina, anterior crus cerebelli, called also *superior cerebellar peduncle* disappears, under the quadrigeminal bodies.

2. Crura cerebelli ad pontem, middle cerebellar peduncles, leave the cerebellum laterally, passing into the pons.

3. Crura cerebelli ad medullam oblongatam or restiform body.

Though the physiological relations are still obscure in many particulars, the following statements may be assumed as probably true.

I. The cerebellum receives its own centripetal tracts, from the periphery of the body, and sends out centrifugal stimulations to the spinal cord.

As centripetal must be named first of all:

1. The lateral cerebellar tract: arises in Clark's column of the spinal cord, and passes through the restiform body into the cerebellum.

2. Gower's tract: arises in the lumbar portion of the spinal cord and loses itself in the cerebellum.

3. Other centripetal stimulations come to the cerebellum from the nucleus of the lateral tract and the crossed olive.

4. Stimulations from the *vestibular nerve* probably by the help of Deiter's nucleus.

By the help of the centripetal tracts, the cerebellum receives messages from the *muscles* of the trunk, neck, head, limbs (unconsciously persisting analog of sensations of position and movement), and from the *labyrinth.*

As centrifugal tracts the following must be first considered:

1. The cerebellar cortex tract (roof nucleus?), Deiter's nucleus, the uncrossed anterior lateral funicular path.

2. The cerebellar cortex-dentate nucleus superior cerebellar peduncle—red nucleus—Monakow's bundle.

3. Connections by means of Deiter's nucleus and posterior longitudinal bundle with the *nuclei for the eye muscles.*

Through these centrifugal pathways muscles are influenced automatically, unconsciously. The cerebellum regulates automatically the holding of the trunk, head, and eyes, especially in standing upright and walking. This regulation of the cerebellum does not suffice, in man, to produce erect

walking; the action of the cerebrum is also essential, but it takes an important part.

The cerebellum represents *first* an independent sense-motor organ, which by its own centripetal tracts (*different* from the sensory tracts, that go to the cerebrum, as the posterior columns, fillet, etc.), receives impressions from the periphery, and by its own centrifugal tracts, makes use of these impressions, by means of the muscles, particularly *for the co-operation of the many factors of equilibrium, that are concerned in standing upright and in walking.*

II. But the cerebellum has (indirect) connections with the cortex of the cerebrum, corticopetal as well as corticofugal. Through the middle cerebellar peduncle, the corticopetal tracts, after various interruptions, reach the *frontal* cortex, from which the corticofugal tracts through the frontal pontile tract and the pontile cerebellar tract reach the opposite hemisphere of the cerebellum.

By means of this cortico-cerebello-cortical system, cerebellar sensations come to be acted on in the cortex of the cerebrum which thus gains influence over the cerebellum. When a frontal lobe is destroyed, the volume of the opposite hemisphere of the cerebellum diminishes.

III. The cerebellum, through its superior peduncles and the red nucleus, sends other stimuli to the cortex of the cerebrum, which this answers, in part at least, through motor processes, *without the assistance of the cerebellum*, by the descending motor tegmental tract, which arises from the red nucleus and ends decussating in the lateral corona.

The main symptoms of disease of the cerebellum and its peduncles, or of the tracts mentioned, are:

1. *Cerebellar ataxia.* Cf. page 506.

2. Subjective: vertigo.

3. Forced postures and forced movements, which are observed, either as an effect of irritation, or as a symptom of loss of function, when the hemispheres of the cerebellum and especially the middle cerebellar peduncle are affected, as well as abnormal inability to move the eyes and nystagmus.

4. The muscles of the body of the same side are usually *hypotonic* (a result of the loss of centripetal stimuli).

5. Occasionally, in diseases of the cerebellum, the ability to carry out antagonistic movements, in immediate succession (as pronation and supination) is lost (diadocokinesis).

6. In contrast to the tabetic the limbs of many patients suffering from diseases of the cerebellum, after a little wavering, may persist for a remarkably long time in one position; at times even *cataleptic* symptoms appear.

7. A species of dysarthria that is occasionally observed (slow, "scanning" speech), could, if not caused by pressure on the bulbar centers of speech, arise from the loss of regulating influences from the cerebellum (analogous to a diadocokinesis).

8. One-sided *chorea* appears in the case of focal injuries that irritate the superior cerebellar peduncle and its connections. A host of other symptoms, observed in diseases of the cerebellum, especially diseases that compress the neighboring organs (tumors), arise from effects on the adjacent pons, medulla oblongata, etc., as hemiplegia, spastic phenomena, vomiting and looking palsy (Blicklaehmung).

Even the direct symptoms of cerebellar diseases are not always present and are often *transitory*, because there may be compensation, in the course of time, for the losses concerned. The cerebrum seems to be able to compensate, to a great extent, for disturbances, that have arisen in the cerebellum. By far the most important and constant symptom is *cerebellar ataxia.*

It is important to note that focal injuries in the cerebellum, as opposed to those in the cerebrum, cause disturbances, in the majority of cases, *on the same side of the body.*

10. Brain and Reflexes

In the first place, the brain has an inhibitory influence on a series of reflexes of the other divisions of the central nervous system. This influence is evinced normally in the possibility of voluntarily checking certain reflexes (coughing, breathing, closing the eyes at the approach of the hand, etc.), furthermore, in the influence exerted on reflexes by mental disturbances (breathing stopped by fear). In lesions of the brain, therefore, by loss of inhibition, many reflexes are exaggerated, such as those of the tendons.

This, of course, is true only so long as a remnant of connection between cerebrum and periphery is retained. When the connection is absolutely broken—total division of the spinal cord—exactly those reflexes belonging to the severed part of the spinal cord are abolished, but in a manner difficult, as yet, to understand. A few authorities still maintain that in cases of a total transverse lesion of the spinal cord above the lumbar swelling the knee-jerks, which at first are abolished, return and are permanently exaggerated.

The brain not only influences spinal reflexes, but is itself the center for certain reflexes.

I. *Pupillary reflexes.*

1. *Light Reflex.*—When light is thrown on the eye, the pupil of that eye contracts (direct light reaction).

At the same time, the pupil of the other eye contracts (consensual light reaction).

2. *Convergence Reaction.*—When looking at near objects (convergence and accommodation reactions) both pupils contract, more than from light. This must be conceived as a concomitant movement.

3. *Orbicular Phenomenon.*—The contracting of the pupils that appears

when the eyes are shut tightly, must also be regarded as a concomitant movement. When this is checked—and a series of concurrent factors does destroy the effect—this contracting becomes evident.

4. *Pupillary Cortical Reflex.*—Contraction of the pupil, from mere direction of the attention to a source of light that has *not* been *fixed*, appears only in few individuals.

5. The asserted contracting of the pupil from a mere *concept* of a bright object, has been proved doubtful by further investigation.

6. *Psycho-reflex.*—On the other hand, without doubt, a *dilatation of the pupils* occurs, not only with all sorts of painful or vigorous stimulations (especially of the skin, but also from noises) but with every psychic disturbance (anxiety, terror) and *energetic mental work or strongly concentrated attention.*

During the vivid conception of a dark object the dilatation of the pupil appears as such a psycho-reflex, which, however, is independent of the content of the concept (anything dark) and the result only of concentrated attention.

In sleep maximum *contraction* of the pupils occurs.

Anatomical

The fibres of both optic nerves end, as we see, half crossed in the lateral geniculate body and the pulvinar. Besides this, fibres reach the anterior quadrigeminal bodies (pupillary fibres). Hence arises a connection with the deeply situated nuclei of the oculomotor nerves, whence a stimulation of the pupil fibres causes contraction of the pupil (Fig. 133).

The tracts of the pupillary light-reflex, as yet not wholly ascertained anatomically (contraction due to light) are as follows:

1. Centripetal limb. Retina, optic nerve, optic tract, anterior quadrigeminal bodies.

2. Central transmission. Anterior quadrigeminal bodies, nucleus of the oculomotor nerve (probably not a direct connection).

3. Centrifugal limb. Nucleus of the oculomotor nerve to the ciliary ganglion and hence with the musculus sphincter pupillæ.

Since in a strong light, both pupils contract (consensual contraction of the unexposed eye), there must be a connection between each anterior quadrigeminal body and *both* oculomotor nuclei.

The *dilatation* of the pupils may result from the innervation of the dilatator pupillæ by the sympathetic nerve.

The center is Budge's *centrum ciliospinale* in the spinal cord, lying at about the level of the origin of the *first* dorsal nerves. It receives its stimulations from the skin through the posterior roots, also from other sources and sends the innervation through rami communicantes to the superior cervical

ganglion of the sympathicus, hence to the Gasserian ganglion, and through the ramus ophthalm. trigem. to the long ciliary branches and to the iris.

But dilatation of the pupils through the dilatator pupillæ, that is through the stimulation of the sympathetic nerve, plays a smaller part than was formerly believed. The dilatation of the pupils in all vigorous sensory irritations, in every psychic exertion and excitement, in concentration of attention, energetic contraction of the muscles, etc., is caused especially by *inhibition of the sphincter center*, i. e., by the oculomotor nerve.

The influence of psychic processes on the dilatation of the pupil—which can easily be proved, for instance, in the case of simple intense recollection by an apparatus for measuring differences—points to connections between the cortex and the sphincter center (Cf), probably also with the ciliospinal center.

Pathology of the Pupillary Reflexes

When a *lesion* destroys any part of the reflex arc, the pupillary reflex is destroyed. The peculiar conditions, under which total loss of pupillary contraction and dilatation, especially when exposed to light, slowness in moving the pupils, hippus, hemianopic reaction of the pupils, myosis, mydriasis, anisocoria (difference in the diameter) appear, are discussed in a special chapter. (See also General Symptomalogy.)

Loss of the psychic-reflex alone, is found frequently in certain mental diseases (juvenile idiocy). The assumption of a so-called "paradoxical" pupillary reaction (dilatation in light), is founded in the majority of cases upon a dilatation that quickly follows an unnoticed contraction.

II. *Eyelid reflexes.*

(a) *Touch reflex:* closure of the eye when the cornea or conjunctiva is touched (trigeminal nerve, nucleus of the facial nerve and this nerve itself). It is absent in diseases of the pons, on both sides also in hysteria.

(b) *Light reflex:* closure of the lid on sudden exposure to strong light, probably caused through the cortex (absent in cortical blindness).

III. *Palatal or choking reflex*, as well as the reflex act of swallowing: sensory divisions in the pneumogastric and glosso-pharyngeal nerves. Center: the nucleus of the motor trigeminus, the pneumogastric, glosso-pharyngeal and hypoglossal nerves. Centrifugal division: these nerves.

IV. *Vomiting.*—Sensory division: pneumogastric, glosso-pharyngeal, splanchnic nerves. Center in the medulla. Motor division: pneumogastric, splanchnic and phrenic nerves.

(a) Coughing and sneezing made possible through the fifth, ninth, tenth, eleventh and twelfth nerves.

We may only suggest the importance of the medulla for the automatic regulation of the heart and respiratory innervations, and of a vaso-constrictor center supposed to exist in the medulla oblongata.

11. Secretory Functions of the Brain

1. For *secreting the sweat*, a center, including the spinal centers, is supposed to be in the medulla oblongata.

2. *Secretion of the Saliva.*—There is in the medulla oblongata a reflex salivary center, which controls the chorda tympani, as well as the sympathetic secretory nerves of the salivary glands (in the passing of the pons into the medulla, in the tegmentum, the so-called nucleus salivatorius). In bulbar paralysis, excessive flow of saliva occurs.

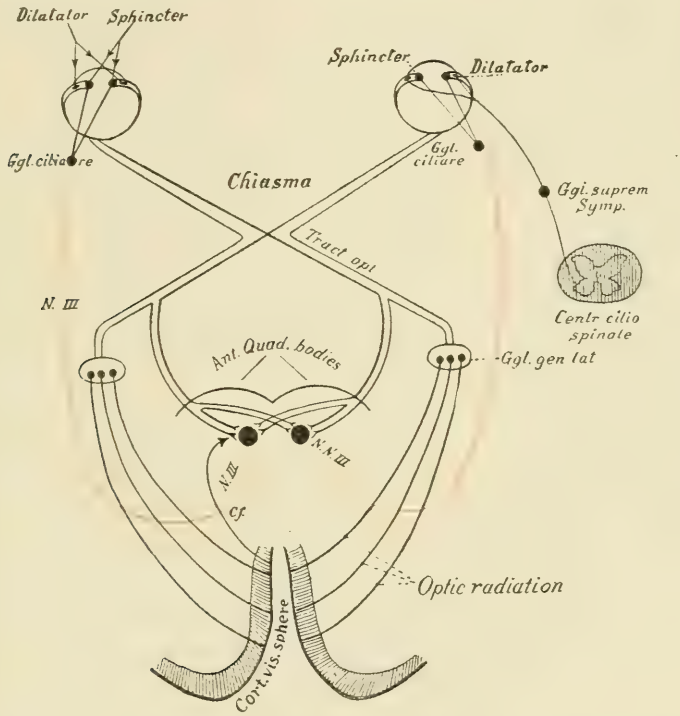


FIG. 133.—Simplified scheme of pupillary reflexes. *Cf*, Presumed centrifugal path from the cortex to the center of the sphincter; *Ggl. gen. lat.*, ganglion geniculatum laterale; *N.N. III.*, nucleus N. oculomot; *N.III.*, nervus oculomot.

Even if the salivary secretion, caused by taste and sensory stimulations of the oral cavity (trigeminous nerve) is *a reflex of the medulla*, the salivary secretion that takes place at the sight, smell, and *imagination* of food (the "mouth waters") must be called a *cortical* reflex.

3. *Secretion of Tears.*—The subcortical reflex has its center in the medulla oblongata. Centripetal fibres run, especially in the trigeminus, and other sensory nerves, centrifugal in the facial nerve. The secretion of tears is under *cortical* influence, as is proved by its dependence on psychic processes.

4. The medulla, through the splanchnic nerves, exerts also an influence on the secretion of the liver (glycosuria in diabetic puncture).

II. LOCAL SYMPTOMATOLOGY

1. General Symptoms

1. *Disturbances of consciousness.*—In the first place there are various degrees of a general diminution of consciousness. In *coma*—for instance, in an epileptic fit, or often, after an apoplectic attack—there is total loss of consciousness.

The reflexes are altogether, or, for the most part, abolished. The limbs, when held up, fall limply down, if left to themselves. The comatose patient can not be roused however strong the stimulus may be.

Slighter degrees of lessening of consciousness are called *sopor* and *somnolence.* The soporose patient is roused by strong stimuli but only for the time being; the somnolent person, is rather easily aroused and capable of reactions, but when left to himself, easily falls back into a drowsy condition, in which most external occurrences have no existence for him.

Associated with these diminutions of consciousness, appear *irritations* of and *changes* in the conscious processes.

These dimmings of consciousness, with their countless *sense delusions*, and the ensuing motor restlessness, are designated as *delirium.* Disturbance of consciousness together with sense delusions causes disorientation. Delirium appears in meningitis, infectious diseases, in the course of alcoholism, morphinism, and also after occlusion and other lesions of vessels.

Anxiousness, and *torpidity* (stupor), even with catatonic phenomena, are observed in organic diseases of the brain. Other mental disturbances, especially general *weakness of memory* (loss of or difficulty in rousing the formerly associated mnemic possessions), disturbance in the *ability to notice* (inability to assimilate anything new into the memory), disturbances in attention, judgment, feeling, volition, supersensitiveness, unnatural suspicion, increased to prejudicial ideas, diminution of ethical motives, narrowing of the circle of interest to the Ego, etc., general symptoms, which appear frequently, as a result of chronic and diffuse disorders of the brain (arteriosclerosis, progressive paralysis, senile atrophy, also multiple sclerosis, tumors, etc.), are described, in more detail, in the text-books of *psychiatry.*

2. *Headache* is a very ambiguous symptom, which, even when the pain is clearly localized by the patient, does not at all warrant the conclusion that the focal injury corresponds to the seat of pain.

3. *Vertigo* a word used by laymen in a very different sense—i. e., for a mere fainting spell or a darkening of the field of vision—means: (1) That the patient's own body or even external objects seem to him to move, at least there is disorientation regarding his posture, or the position of the body itself, that is, *subjective vertigo* or (2), loss of equilibrium, that is, *objective vertigo.* When severe the patient may fall. Vertigo, especially

if slight or if subjective, is seen in several different conditions, i. e., anæmia of the brain, general vascular changes, cerebral hemorrhage, etc. But the higher degrees stand in particular relation to *cerebellar* and labyrinthine diseases. (Cf. pages 481 and 15.)

4. *Vomiting* is a symptom of local irritation in the medulla oblongata, and is classed among the general symptoms only because this center is easily stimulated to reaction by *pressure* and *irritation* in *far distant* parts of the brain, or perhaps reflexly, by disturbances of the sensory branches of the dura.

Cerebral vomiting is distinctly characterized by its occurring without nausea, the ease with which it occurs, and its occurrence when no nourishment had been previously taken. It is often projectile.

5. Changes in the *pulse, respiration, temperature.*

Cerebral diseases can influence, in many ways, the action of the heart.

Slowing of the pulse, which frequently appears when there is a rapid increase of pressure within the cavity of the cranium, by irritation of the pneumogastric center in the medulla oblongata is of special importance.

Even in the absence of an infectious disease, a transient rise in temperature occurs after arterial occlusion, or cerebral hemorrhage or a paralytic seizure, without any certainty whatever, as to what definite parts of the brain are directly concerned. On the other hand, the temperature occasionally falls to 96 degrees.

Of the many changes from normal respiration, which appear especially in coma, the most important is the *Cheyne-Stokes'* respiration, which occurs also in the final stages of heart and lung diseases and uræmia, and is a "signum pessimum." It is marked by the following series of irregularly interchanging phases: 1. A number (20–30) of increasingly superficial breaths, following one another in quick succession. 2. Pause in breathing. 3. Little by little breathing beginning again, at first slow and deep, but gradually increasing in rapidity till phase 1 is reached. This sequence recurs again and again.

6. *Choked disc* (merely a severe optic neuritis) presents ill-defined boundaries of the turgid, reddish, and swollen papilla, with total disappearance of its boundary, often with hemorrhage and white spots, enlarged and tortuous veins, and constricted arteries; the vessels seem to be broken off at the edge of the disc. Choked disc is a general symptom, because it occurs in consequence of any space-restricting lesion anywhere within the cranium (especially in *tumors*, internal hydrocephalus, abscesses), also in meningitis, less often in cerebral syphilis and lead poisoning and very seldom in multiple sclerosis. It occurs most frequently with tumors of the *posterior* cranial fossa. It is most frequently *bilateral*, even when appearing, as it often does, on one side first.

2. Projection Disorders

Disorders in Motion and Sensation

We differentiate:

1. *Projection Disorders.*—They affect *movement, sensation,* and *secretion. Phenomena of irritation or loss of functions* of receptor or effector organs.

2. *Mnemic-associative Disturbances.*—They affect those functions of the brain which consist in *storing up* past stimulations, and in fixing their *connection.* The associative work of the brain does not correspond to any arrangements of the sense and muscle apparatus of the body, but to the connections established by *experience;* so, for instance, the association between the scent and the appearance of the rose, is not at all brought about by the connection between the mucous membrane of the nose and the retina.

The projection disturbances are divided into motor, sensory and secretory, and each of these groups, into symptoms of *irritation and of loss.*

(a) Motor Symptoms of Irritation

Cortical spasms. As the electric current, and all other irritants that influence the motor cortical centers, cause contractions in the muscles of the limbs on the other side of the body, so with pathological irritants (tumors, hemorrhage, scars, etc.), spasms occur in the limbs. Usually the spasm is first *tonic,* later *clonic.*

According to the place in the cortex that is affected by the irritation, the muscles of the face, arm, or leg are the seat of the spasm. When the irritation is lasting and extended, the spasm spreads from the region first attacked to the other limb on the opposite side and it proceeds in such a way, *that the muscles are affected, in the order in which their cortical centers are arranged,* cf. page 475, so that the spasm that has started in the face, continues first in the upper, then in the lower extremities. Finally, the spasm can pass over *to the same side* of the body, in which the focal injury lies, whereby the clinical picture of a general epileptic fit is produced. Even when the spasm does not, *universally,* attack the other side, the *usual bilateral* muscle groups (of the jaw, for breathing, etc.—viz., in what is otherwise one-sided spasm) are also affected by twitching, even though they lie on the other side.

These cortical spasms, confined to one group of muscles, or to one side of the body, or at least, beginning locally, are known as *Jacksonian* or *cortical* epilepsy. They are contrasted with the convulsions of ordinary epilepsy, which affect the whole body from the start.

After severe Jacksonian convulsions, paresis appears in the muscles attacked, as a consequence of exhaustion, but retrogresses after a few hours, at most. But if in the progress of the disease, as often happens, though the effect of irritation is only temporary, the lesion causing the spasms, e. g., a

neoplasm leads to *destruction* of the motor cortical center, permanent paralysis of those groups of muscles, which were formerly affected by the spasm, appears. Together with the extremities, on the opposite sides, head and eyes fall into a mostly tonic spasm. If the irritating focal injury lies towards the left, both eyes and the head turn towards the *right:* conjugate deviation (déviation conjugué). In this deviation, produced by an *irritative* cause, the patient looks towards the side on which the limbs are spastic, contrary to the deviation caused by paralysis. It is worthy of notice, that not only the contralateral eye, but both eyes are affected by the effect of this irritation which exists in one hemisphere alone.

This combination of tonic-clonic spasms in isolated groups of muscles on one side, can occur, pathologically, only with cortical irritation. It must be remembered, however, *that focal injury not immediately in the center concerned, but adjacent to it, can cause the irritation.*

Therefore spasm is of much less importance in the diagnosis of the localization of disease than is palsy. *Symptoms of lost function are of much greater surety, for local diagnosis, than those of irritation.*

General spasms, and especially, tonic spasms can also be discharged from deeper lying subcortical centers. In irritation of cortical motor centers (Jacksonian epilepsy), the characteristics are: spasmodic contractions, confined to a group of muscles, or at most, to one side, the change from tonic to clonic, and its spreading according to the location of the centers for individual limbs, in the anterior central convolution.

Choreic Twitchings. Hemichorea

Movements of the limbs much resembling those of Saint Vitus dance in children and parturient women, appear in focal injuries, *on one side,* in paretic limbs (often long after the paralysis), or sometimes precede, by a considerable time a paresis or paralysis. (*Hemichorea.*) If, as is usual, they appear in hemiplegic limbs, we have posthemiplegic chorea. The affected limbs are often hypotonic. The hemichorea is sometimes accompanied by pain. The kind of disturbance in movement varies in different cases, sometimes resembling mere tremor, at others athetosis.

As to the anatomical basis of hemichorea, as a rule focal injuries in the subthalamic region (red nucleus and its connections with the cerebellum (superior cerebellar peduncle) and the optic thalamus) seem to be the cause.

Athetosis, a mostly *unilateral,* unceasing, involuntary movement, appearing in paralyzed limbs, abating only in sleep. It consists in slow spreading, adducting, flexing, and extending movements, especially of the fingers and toes. (Athetosis of the facial muscles is of rare occurrence.) Over-extension of the fingers is conspicuous. Athetosis appears most frequently in cerebral infantile paralysis, but not until long after the onset

of the paralysis, and when it has, to a certain extent, retrogressed. Athetosis, too, seems to be caused by focal injuries in the sensory tracts leading to the thalamus. It may appear, exceptionally, with diplegic paralysis, in both halves of the body.

The so-called double athetosis (Athétose double) is a disease sui generis, which appears symmetrically on both sides without preceding paralysis from an unknown cause and without anatomic concomitants.

Unilateral tremor is found occasionally in hemiparetic limbs. In many cases it resembles perfectly the marked tremor of *paralysis agitans*.

Clonic-tonic twitchings of the palatal muscles, 150 to 200 in the minute, have been observed, especially with pseudo-bulbar symptoms, in hemiplegic patients, and seem to have been traced back to injuries in the vicinity of the red nucleus.

Associated movements are involuntary movements appearing in other limbs or in other muscles of the same limb while this limb is being moved purposely or even moving reflexly.

They appear especially in the paralyzed limbs, when strong movements occur in the sound limbs. But also in the sound limbs, when an attempt is made to innervate the paretic ones. Thus, when the sound hand is energetically closed, an impulse discharges itself into the paralyzed hand, which can not voluntarily be closed, and it closes or an unsuccessful attempt to close the paralyzed hand has the effect of making the sound hand into a fist. With sneezing and yawning a paralyzed limb sometimes extends or flexes.

Strümpell's tibialis phenomenon; strong dorsal flexion of the foot while the hemiplegic leg is being drawn toward the body, is an associated movement of another group of muscles of the same limb. Even in passive movements of the sound limb, the paretic limb may sometimes carry on similar movements.

In paralytics, chronic alcoholics, idiots, and aphasics, we often find, when they are speaking associated movements of the facial muscles, especially of the forehead.

Contractures will be discussed later on with the paralyses. See page 493.

(b) Sensory Irritation Symptoms

In focal injuries, that irritate the sensory tracts (see below), constant, or intermittent violent pains may appear, which, to distinguish them from the usual pains produced by peripheral irritation, are called "*central pains*." These pains, sometimes combined with a feeling of heat, are felt in the opposite side of the body, or it may be, too, in a single limb, arm, or in the face, tongue, etc. Besides pains, paræsthesias of various kinds, formication,

thermic sensations, etc., may appear. Unilateral sensations of cold of sudden onset have been noticed, and sensations of movement, without objective movement.

Sensibility may remain undisturbed in the presence of these central pains; but pains may appear in regions insensitive to external irritation—anæsthesia dolorosa.

Sometimes, hemichoreic twitchings, produced by focal injuries, are combined with central pains.

As irritation phenomena, we may regard the *hyperæsthesias* also: even a slight touch, or a slight thermic stimulus is painful, as well as the *paræsthesias* due to touch alone: strange, "funny" sensations.

Central pains and paræsthesias may be caused by focal injuries in every part of the sensory system, even in the cortex and corona radiata, more frequently by those in the *thalamus* and *subthalamic region.* Therefore they have no localizing value.

In the territory of the *special senses* (sight, hearing, etc.), stimulation phenomena appear in the form of elementary sensations, the seeing of light, flames, colors, the hearing of humming, ringing and rustling, and of complex *hallucinations.* Thus disease in the occipital lobe, may cause visual hallucinations in the hemianopic half of the field of vision.

(c) Motor Symptoms of Loss of Function

1. *Paralyses*

Soon after a grave apoplectic seizure, the so-called *initial flaccid paralysis* appears; the passively raised limbs fall as if lifeless.

The reflexes are lost at first, but soon a certain tonus appears in the muscles, the tendon reflexes return and are soon exaggerated, the Babinski plantar reflex appears. The stage of flaccid paralysis usually lasts only a few days, rarely, a few weeks. In some muscles a certain degree of motility returns, e. g., in those which flex the arm and fingers, and now gradually the flaccid paralysis passes over into the permanent condition of residuary hemiplegia, to be described immediately.

In residuary hemiplegia, it is not, as after complete division of nerves, a question of paralysis in the strictest sense. The muscles are not as in the case of nerve division robbed of all tonicity, nor are they incapable of reflex movements. On the contrary, in part of the muscles hypertonia appears and the tendon reflexes are exaggerated.

The behavior of the *reflexes*, discussed on page 64, will be only touched upon here.

In cerebral paralyses a few days after the attack we find exaggeration of the tendon reflexes and mechanical muscular irritability. Often *patellar and ankle clonus*, less frequently hand and masseter clonus are found. Tapping on many other tendons (periosteal spots) and muscles, results in in-

creased contractions (supinator, triceps, radius-periosteal reflexes, etc.). In a certain but not in absolute contrast to the tendon reflexes stand certain skin reflexes. The abdominal and cremasteric reflexes are generally abolished on the paralyzed side. But one must consider, that, as a matter of fact, even the former can not always be obtained in healthy people, and that therefore, only a one-sided loss is significant. (In long-standing hemiplegia the knee-jerk may be greatly increased on the unparalyzed side.)

If exaggeration of the reflexes should, exceptionally, fail to appear, either a concomitant disease of the posterior column (tabes) or neuritis is the cause.

A frequent symptom—in 70–80% of cerebral paralyses—is the *Babinski* plantar phenomenon (see page 69). *Oppenheim's* dorsal leg phenomenon (see page 69) and *Kurt Mendel's* reflex (plantar flexion of the toes instead of the normal dorsal flexion, when the dorsum of the foot is tapped, near the third and fourth metatarsal bones) are frequent signs of paralysis caused by lesion in the cortico-spinal path.

In the hemiplegic limbs, all muscles, to a certain degree, are weakened, but some groups of muscles, the so-called *preferred* muscles of hemiplegia remain strongly affected while the others recover somewhat. These preferred muscles belong to definite mechanisms. The peculiarity of all cerebral paralyses, is, that not single muscles, but entire muscular *mechanisms*, i. e., many muscles working together, called *synergies*, are paralyzed. One must consider, that a simple movement (such as extending the index finger) is not the work of a single muscle. Antagonists are involved as well as collateral and rotatory synergists.

The mechanisms particularly injured in cerebral paralysis are, in regard to the leg, all the mechanisms which help to *retract* the leg, especially those which bend the knee and the dorsal flexors of the foot. Because of the weakness of the dorsal flexion, the tip of the foot often drags on the ground and the leg is lengthened. The *extensors* of the leg are retained, especially those of the leg below the knee and the plantar flexors of the foot. Therefore, the leg may be used stilt-like in walking. In any case, the leg is less seriously affected than the arm. The glutæus medius is often paralyzed, which fixes the pelvis against the thigh. Consequently in walking, the *pelvis* falls towards the unparalyzed side. In order to compensate for this, the hemiplegic patient, every time he steps on the paralyzed side, throws his trunk towards that side. Hence occurs the peculiar sideward movement of the trunk in walking, found in many hemiplegics. In double paralysis of the gluteus medius—a rare phenomenon of cerebral infantile paralysis—a waddling walk results. The body, at every step, sinks to the side of the standing leg.

In the upper extremities, the muscles that open the hand, and rotate it and the arm outward, are more seriously paralyzed, while those of opposite action, closing the hand, and inward rotation of the arm, are less seriously affected.

The raising of the shoulder (M. cucullaris and levator scapulæ) is seriously affected. The *sternocleido-mastoid muscle, on the contrary, is always unaffected.* For both upper and lower extremities, this rule holds good, that the *distal* parts are always more seriously affected, i. e., hand and foot, and *especially the differentiated movements of fingers and toes, the isolated movements in the small joints.* In contrast to these, there is retained ability to make a few gross changes in the position of the entire limb.

The frontal ocular branch of the facial nerve is spared (usually, however, the eye on the paralyzed side can no longer be closed alone), whereas *the branch running to the mouth and cheek is clearly paretic.* (An important difference from the peripheral paralysis of the seventh nerve.) The mouth is drawn to the unaffected side.

The *tongue*, when stuck out, turns to the paralyzed side, because the sound geniohyoglossus presses to the other side, but for the rest, is sufficiently mobile.

Symmetrically working muscles, on the two sides of the body, which, as is well known, may be innervated by each hemisphere individually (muscles of the eye, back, larynx, of chewing and swallowing) are unaffected or almost unaffected in hemiplegia.

The bilaterally working speech muscles, too (of the tongue, palate, face), are not so deprived of function by one-sided focal injury, that speech is permanently lost. Serious dysarthria, for the most part, appears only as a transitory phenomenon, after one-sided injuries and only when they are left-sided. Lasting anarthria or serious lasting dysarthria, appear almost only in *bilateral focal injuries*, in contrast to *aphasia*, which appears only after lesions on the left side.

2. *Contractures and Hypertonia*

A further factor in restricting motion (myogenic contracture) is added to actual muscular weakness in hemiplegia, after the initial flaccid palsy has passed off.

By this one understands the fixation of limbs in a definite position by the occurrence of involuntary permanent shortening of muscles.

These myogenic contractures are to be distinguished from the contractures due to cutaneous scars, ankylosis, or shortening of ligaments or tendons.

The normal tension of the muscles, fixes in a certain measure, in healthy conditions, the limbs in their positions at all times, so that in sudden and quick passive movement of a limb, a moderate resistance of the stretched muscle is felt. This normal resistance is lacking in some nervous diseases, especially in tabes. Then we speak of increased passive motility or of hypotonia.

On the contrary, the resistance to passive movement, in diseases of the pyramidal tracts, is very much increased: *hypertonia* of certain muscles and, therefore, fixation in definite positions appears.

In this fixation, there is a second factor; frequently there appears besides hypertonia, *connective-tissue contraction. Contraction of the connective tissue* of a muscle appears, when its points of insertion are not separated from one another for a long time, therefore, in the antagonists of a paralyzed muscle, *even if* it is not in the condition of hypertonia. Shrinking contracture, therefore, appears also in *flaccid* paralyses, as, for instance, in poliomyelitis.

In *spastic* paralyses, which have existed for a long time, there *is added* this shrinking contracture to the hypertonic or spastic contractures.

These *shrinking* contractures must, therefore, be differentiated from the spastic, which are conditioned not by tissue changes, but by nervous hypertonia. This *hypertonia* exhibits itself in hemiparetic limbs, even when it does not go so far as contracture, by increased spring-like resistance on *sudden passive flexion or extension*, and corresponds to the spasm, which appears with the patient's own movements. The reflexes are exaggerated.

Sometimes one sees, either at once or soon after a cerebral injury occurs (hæmatoma of the dura or trauma, meningitis), *spasmodic*, rigid, abnormal attitudes and positions of the limbs, which, unfortunately, have been called early contractures. They usually disappear again and alternate with tonic-clonic spasms. They are conditions of irritation, better included under tonic spasms. If one speaks broadly of contractures, one does not mean these transient increases of tension caused by special irritation, but permanent conditions of contracture, which appear as a permanent symptom in from a few days to one or two weeks, after apoplexy, resulting from a lesion in any part of the entire motor cortico-spinal system, that is, from the cortex of the anterior cerebral convolution to, but not including, the anterior cornu cells.

Unless therapeutic preventative measures are taken in these cerebro-spinal paralyses, a shrinking contracture in the course of time is added to the spastic contracture.

Spastic contractures are relaxed in *narcosis*, in deep *sleep*, under the effect of constriction in *Esmarch's bandaging of the affected limb*, and also partly in a warm bath.

Naturally shrinking contracture can not, under these circumstances, be relaxed, and thereby the part that hypotonia and shrinking play in contracture, can easily be determined. Mental excitement increases spastic contracture. Spastic contracture shows a spring-like resistance against attempts to cause relaxation. It is violent in repelling sudden brusque attempts; on the other hand, many contractures can be easily relaxed by *gradual* procedure. Attempts at movement both in the affected and the unaffected limbs, increase the contracture.

The characteristic positions of the limbs of patients suffering from mono-

hemi-, and diplegia, appear only because of differences in the degree of contracture in the various groups of muscles.

There is, indeed, absolutely no regular form of hemiplegic contracture, but one type is the most frequent. On the whole, *we find contracture in those muscles that are relatively free from paralysis* (cf. page 492), that is, the antagonists of the "predilection" muscles. The typical position of the hemiplegic arm corresponds, therefore, to the power retained by the flexors of the hand, the inner rotators and flexors of the arm: the fingers are closed, the thumb turned in or adducted, the arm lies close to the breast, bent at the elbow, the leg in extension, the foot in the position of equino-varus. Because of the lengthening of the leg by the plantar contracture or weakness of dorsal flexion, the leg in walking is moved in a circle (circumducted). This is the usual distribution of contractures, in which generally, the muscles that may still be somewhat moved at will, are contracted. Sometimes, but more seldom the opposite takes place: flexion and contraction of the leg, extension and contraction of the arm and fingers.

The distribution of contractures and the corresponding permanent position of the paralyzed limbs, certainly depend on the fact that certain muscles are relatively spared. But only *indirectly*, through the fact, that the return of a certain power in single groups of muscles *is decisive for the position of the limb in the first weeks after the paralysis occurs.* In the final issue, the position assumed by the limb during the time of relaxed and semi-relaxed paralysis determines the distribution of contractures. Each group of muscles is inclined in pyramidal disease to adapt itself to an approach of the insertion points, brought about by any cause whatsoever, through increase in tension and shortening. This is a subcortical reflex, which appears only after the "isolation" of the subcortical centers (and besides depends on the intactness of centripetal pathways, hence is missing when tabes exists).

That this assumption of the dependence of contractures on the position taken by the limb after paralysis, is correct, is proved by the fact that one is able, by artificial fixation of a limb, to change and transform an already existing flexion contracture into an extension contracture and vice versa.

The return of power to certain groups of muscles (the antagonists of the preferred (predilection) muscles) seems, therefore, to lead to contracture only by causing the choice of a certain position of the limbs in the first weeks after the paralysis. Hence it happens that, if, by external conditions, the limb is prevented from following this influence, the distribution of contracture is changed, and in this way mainly can we account for the various modifications of the above described *typical* position of the limbs in hemiplegia.

(3) *Differences in Cerebral Paralysis According to the Situation of the Lesion*

(α) *Cortical Focal Lesions*

The fact that the motor paths in the inner capsule and the cerebral peduncle are placed closely side by side, on the one hand, and the fact that

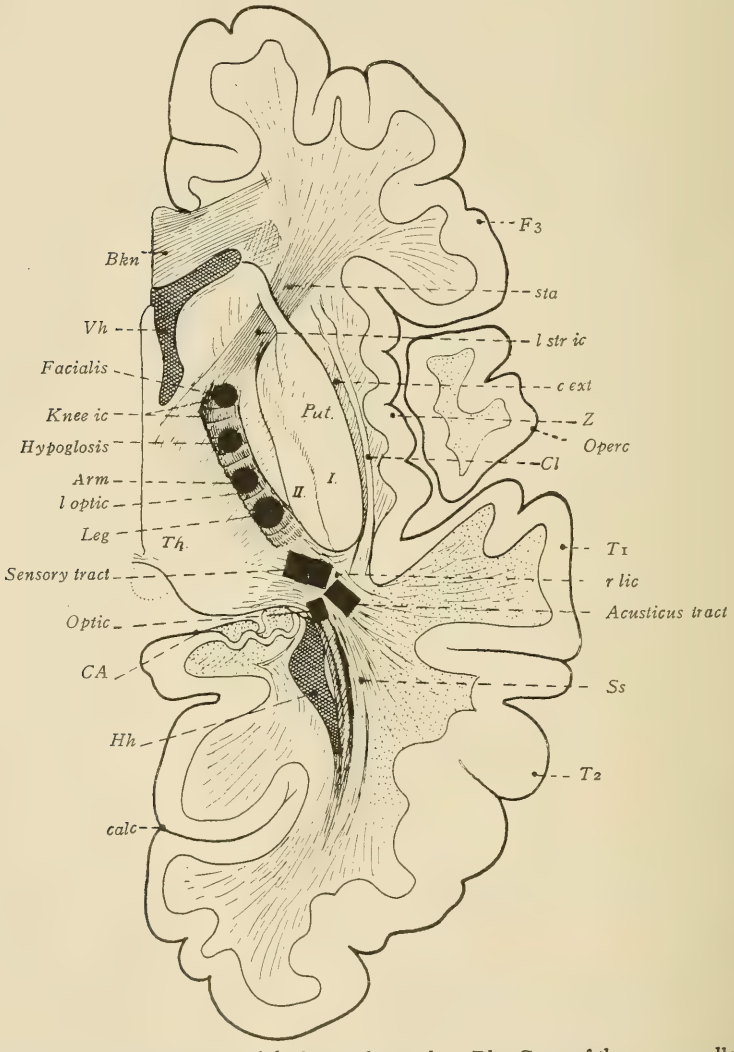


FIG. 134.—The principal segments of the internal capsule. *Bkn*, Genu of the corpus callosum; *Vh*, anterior horn; *Knee ic*, genu of the inner capsule; *l optic*, lenticulo-optical section of the internal capsule; *CA*, cornu ammoni; *Hh*, posterior horn; *calc*, fissura calcarina; *F*3, third frontal convolution; *sta*, corona radiata; *l str ic*, lenticulo-striate section of the internal capsule; *c ext*, capsula externa; *Z*, island of Reil; *Operc*, operculum; *Cl*, claustrum; *T*1, first temporal convolution; *r lic*, retro-lenticular section of the internal capsule; *Ss*, optic radiations; *T*2, second temporal convolution. (*After v. Monakow.*)

they are separated in the white matter, corresponding to the various parts of the anterior central convolution, from which they arise, on the other hand,

cause a difference in the effects of focal injuries, that affect the cortex and parts of the adjacent white matter from those focal injuries affecting the inner capsule, or the cerebral peduncle, and, of course, the still deeper lying planes of the motor path. The paths for the leg come from the paracentral lobule and the upper fourth of the anterior central convolution, those for the arm and hand from the middle two-fourths, those for the face, tongue, masticatory muscles, from the lower fourth of the same convolution.

Focal injuries in or near the cortex, of moderate extent, paralyze *only* arm or leg or face, that is, cause so-called *monoplegias*, whereas focal injuries more deeply situated paralyze most frequently the entire contralateral side, causing *hemiplegia*.

Monoplegia of the face is often combined with that of the arm: monoplegia facio-linguo-brachialis; that of the leg, often with that of the arm, monoplegia brachio-cruralis.

A lesion entirely in the paracentral lobule causes merely *paralysis of the leg*. As the paracentral lobule is supplied by another artery than the central convolution, namely the *corpus callosum artery*, this isolated paralysis, combined with *softening of the corpus callosum* occurs when the above-named artery is occluded.

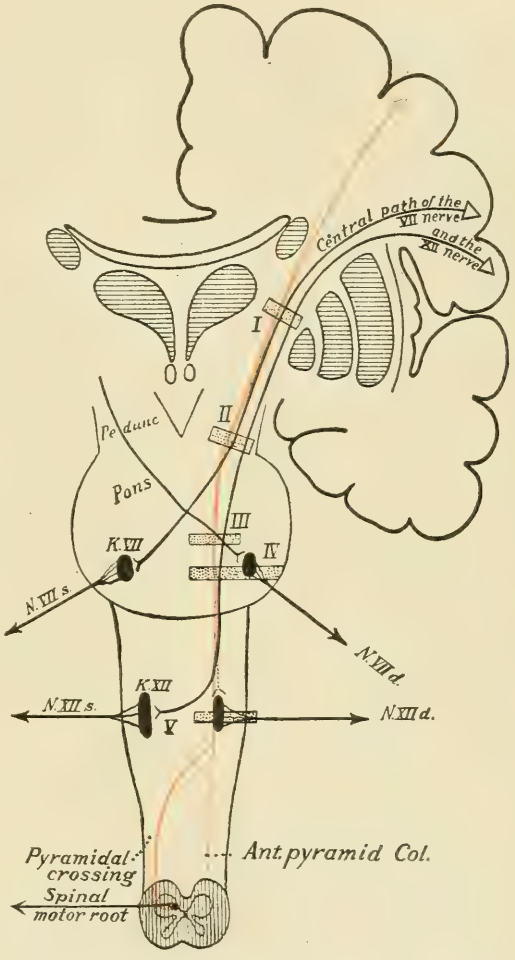


FIG. 135.—Scheme for the explanation of hemiplegia alternans (for the seventh and twelfth nerves). Red: pyramidal tract. *N.VIIs*, Nerv. facialis sinister; *N. XIId*, right hypoglossal nerve. *I*, Focus in the right internal capsule, left half of the body, left seventh and twelfth nerves affected; *II*, in the right peduncle, the same; *III*, focus in the right half of the pons, left half of the body and left twelfth nerve, right seventh nerve (supranuclearly), affected; *IV*, in the same place caudad, left half of the body, left half of the tongue and right seventh nerve affected (nuclearly resp. radicularly); *V*, focus in the medulla paralyzes the left half of the body and the right half of the tongue (nuclearly or radicularly).

Therefore the characteristics of cortical focal injuries, or of focal injuries lying near the cortex are:

1. The paralysis is confined to one or two members (arm, leg, face).

2. Localized Jacksonian convulsions.

In lesions of the central convolutions, inner capsule, etc., the muscles of the eye are not affected. For conjugate deviation see page 478.

(β) *Capsular Focal Lesions*

In the internal capsule, all the motor paths are easily stricken together; they lie back of the so-called knee in the front third of the posterior limb.

The order of fibre bundles, from the knee backwards, is as follows:

Facial fibres, tongue fibres, arm fibres, leg fibres (Fig. 134). The fibres are so indistinctly separated that a monoplegic paralysis through capsular focal injury is exceedingly rare. It has been observed that the leg is *mostly* affected by an injury situated in the hind part, the face by one in the front.

Focal injuries in the cortex and the white matter differ in one respect, that has long been disregarded, from those in the inner capsule and still deeper lying parts of the pyramidal tract. The former affect the fibrillation of the hemisphere, where it contains closely mixed, projection fibres and corpus callosum fibres, destroy therefore besides these, countless connections with the other hemisphere. On the contrary, focal injuries in the inner capsule, in the peduncle, etc., destroy no commissural fibres and therefore leave the harmonious co-operation of the two hemispheres undisturbed. A consequence of this difference we shall learn later on under *apraxia*.

(γ) *Focal Lesions in the Peduncle and Pons. Alternate Paralyses*

If a focal injury affects the motor path in the *peduncle*, where it is situated in the middle third of its foot, the result for the extremities is the same as if there were a break in the inner capsule.

But for the cerebral nerves here and in the *pons*, certain peculiarities occur. Whereas the pyramidal tracts for the extremities do not cross until the medulla is reached, the corresponding paths for the cerebral nerves pass already in the peduncular layers or pontile layers to the nuclei of the nerves concerned on the other side (third, fourth, fifth, sixth and seventh nerves). Such a focal injury in the left peduncle may strike the entire cortico-spinal path for the right extremities, and also the nucleus or root of the left oculomotor nerve. Consequently to the spastic paralysis of the right limbs and the right seventh nerve, is added a nuclear or peripheral paralysis of the left oculomotor nerve (ptosis, internus, paralysis, etc.). This is called *hemiplegia alternans* (alternate hemiplegia).

The same thing, moreover, can take place in the pons for the trigeminus, abducens, and facial nerves. Then to the paralysis of the limbs of one side is added paralysis of the trigeminus, abducens or facial nerve, on the other

side, and moreover, when nucleus or root is affected, the paralysis is of a peripheral character. It may happen, however, that only the supranuclear bundles of the central neuron are affected.

Because of the relation of the nuclei to the pyramidal tracts, many combinations are possible. For instance, when the focal injury crosses the middle line, both seventh nerves or both twelfth nerves may be stricken by supranuclear paralysis, or one by supranuclear and the other by nuclear paralysis besides a hemiplegia (Fig. 135).

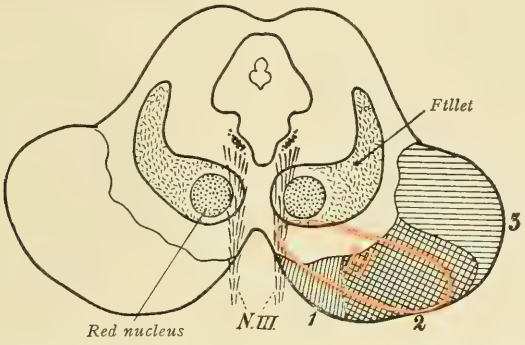


FIG. 136.—Hemiplegia alternans superior. The focus *H* interrupts the right pyramidal tract 2 and the right oculomotor root *N.III* and produces therefore paralysis of the left extremities and of the right eye.

The combination of hemiplegia with crossed oculomotor paralysis (peduncular focal injury) is known as Weber's paralysis or hemiplegia alternans *superior*.

Hemiplegia with crossed paralysis of the facial or abducens nerve must result from a focal injury in the pons (Gubler's paralysis). Hemiplegia alternans inferior.

In the medulla, a focal injury may strike the as yet uncrossed pyramidal tract of the one side, and the root of the hypoglossal nerve of the same side; then the limbs are spastically paralyzed on one side and the tongue atrophically paralyzed on the other (hemiplegia alternans infima).

Just as cerebral motor nerves crossed to the extremities, can be paralyzed by focal injuries in the peduncle and pons, there can be *disturbances in the crossed sensory nerves*, of such kind, that with the pyramidal tract of the right half of the pons, the right root of the trigeminal nerve is affected, hence with paralysis of the left side of the body, disturbances in sensibility in the right side of the face may appear (Fig. 137).

These *interchanging* paralyses of the cerebral nerves are *important local symptoms of lesions in the peduncle and pons.*

Volume of Muscles

In contradistinction to nuclear and peripheral paralyses, the muscles in cerebral paralysis retain, at first, their volume and gradually atrophy at a

rate corresponding to their remaining unused. Atrophy is not so marked as in nuclear and peripheral palsy. Likewise electric reaction is not qualitatively changed in muscles and nerves, and quantitatively is but slightly lowered. Exceptions are rare (atrophy and electric changes). Their cause is not known.

Vasomotor and *trophic* disturbances are observed in hemiplegic and sometimes in merely hemianæsthetic limbs; changes in temperature, cyanosis, edema, joint diseases, skin diseases, etc., also occur.

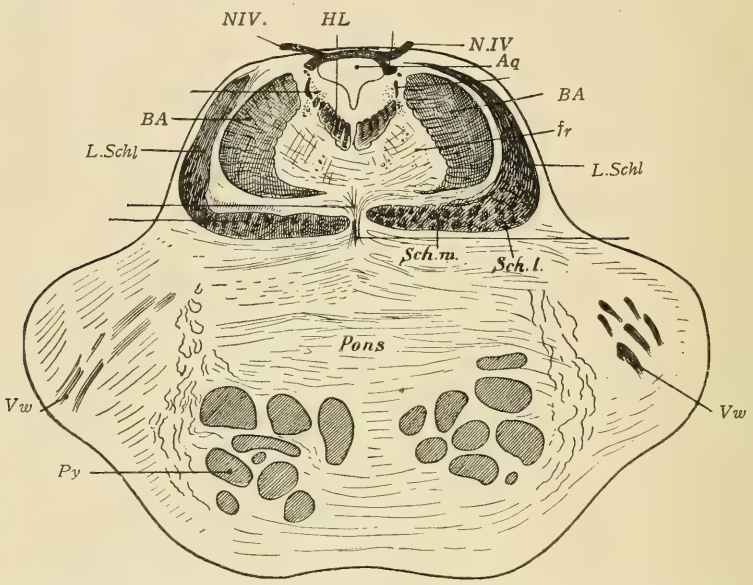


FIG. 137.—Relation of the trigeminal to the pyramidal tract in the pons. A focus, which strikes the root of the fifth nerve and the pyramidal tract *Py*, produces synchronous paralysis of sensibility in the face and crossed hemiplegia. *Sch.m* and *Sch.l.*, middle and lateral parts of the principal fillet; *L.Schl*, lateral or lower fillet; *fr*, formatio reticularis; *BA*, superior peduncle; *HL*, posterior longitudinal bundle; *N.IV*, trochlearis, *Vw*, root of the trigeminal. (*After v. Monakow.*)

Bilateral focal injuries cause bilateral hemiplegia—*diplegia*. Focal injuries that strike the paracentral lobules only, paralyze *both* legs (cerebral *paraplegia*).

(d) Sensory Symptoms of Lost Function

General sensation includes various qualities:

1. Tactile sensations.[1]
2. Sensations of pain (loss: analgesia or hypalgesia).
3. Thermic sensations (loss: thermoanæsthesia).
4. Sensations of place (localization of the place of stimulation).

} Superficial sensibility.

[1] Strümpell differentiates pressure sensations of the deeper parts.

5. Sensations of space. Differentiation of two simultaneously stimulated points on the skin as separate (Weber's circles of touch).

6. Sensations of situation and of movement, the latter both passive and active.
7. Sensations of weight and resistance.

} Deep sensibility of joints, tendons, muscles (also called muscular sense).

A "stereognostic" sense, that is the ability to *recognize* the form of a body from sensations of touch, position, movement, obtained by handling it, may not be placed *with* the *elementary qualities*.

Astereognosis, in so far as it is not the consequence of the loss of elementary sensations, belongs among *agnostic* symptoms. (See page 541.)

A loss of sensation may be partial, in so far as only certain qualities are lost, for instance only sensations of position and movement (*dissociation* of sensibility).

It can also be partial in relation to its *intensity* (loss, mere diminution of sensibility).

Finally in relation to its *extent*. Only part of half of the body, or only part of a member is anæsthetic or hypæsthetic.

In order to cause complete loss of all sensation in the distribution of any nerve, either the nerve or its root must be completely cut off. Even then partial compensation may occur in the course of time because of the partial overlapping of the sensory areas by various roots.

Such total anæsthesia can be caused by cerebral focal injuries only in the pons or medulla oblongata, since only in these places can the peripheral neuron of a nerve which subserves general sensibility, namely, the root of the trigeminus be affected. More centrally, a supranuclear central neuron is always struck by the focal injury. Lesions of the central sensory pathways, in whatever plane they may lie, usually do not *totally* destroy sensibility in any of the three directions (quality, intensity, extent).

Anæsthesias of the entire half of the body, contralateral to the injury, endure only for the first days or at most weeks after the attack.

In particular, sensibility to pain is nearly always to a certain extent restored, even if it frequently remain diminished.

Next *thermic* sensibility is most frequently restored, and after it sensibility to *touch*. Sensations of *place* and *space*, *position* and *movement*, on the contrary, in cases of central focal injuries, are often permanently destroyed.

On the whole, even in large cerebral focal injuries, sensory processes are better restored than motor processes.

Concerning the extent of sensorial disturbances, it must be noticed, that at

first they frequently exhibit an hemianæsthetic character (the anæsthesia ceases precisely at the middle line) but that later the boundary recedes from the middle line—the medial parts of the head and neck again become sensitive—and the disturbance becomes less from the *distal* parts of the body, hand, foot, etc.

Hemianæsthesia, as long as it is total, reaches the middle line and affects not only the skin, but the mucous membranes, joints and muscles. It is accompanied usually by hemiplegia, but there are also cases of isolated and nearly isolated cerebral sensory disturbances.

Anæsthesia may also confine itself wholly or approximately to single limbs or parts of limbs. (In that case, the distal parts are more seriously affected.)

Generally the more seriously paralyzed limb, is also more seriously impaired in respect to its sensory functions.

The dissociation of sensibility by cerebral lesions may in exceptional cases go so far, that cold sensations are retained, while heat sensations are lost.

Concerning *ataxia* of the injured side, in so far as it is connected with disturbances in muscular sense, see next division.

The above general statements concerning the usual *incomplete* loss of sensibility, or, at least, the probable restoration of part of the sensibility, holds good first of all, for the *cortical* parts and those lying near the *cortex*, but to a certain degree also for focal injuries in the thalamus, even for those in the pons and medulla.

The sensory pathways obviously have manifold *lateral branches*—as is proved by their frequent interruption by gray matter. One must consider even the vicarious entrance of the other hemisphere, since in some cases return of a considerable amount of sensibility has been observed, though all the sensory paths of one hemisphere had been *totally* destroyed. Gradual processes, like tumors, cause far less sensorial disturbances, than those appearing suddenly.

Peculiarities of focal injuries at various levels must be discussed under a review of the *sensory paths*.

Our knowledge of the sensory pathways is, as yet, very incomplete; opinions vary greatly on the more delicate relations of parts; therefore the following statement can give only a rough *schematic* sketch.

The centripetal paths, that course to the *cerebellum*, and do not cause stimulation that leads to conscious perception (*lateral cerebellar tract and Gower's tract*) have been mentioned in the discussion of the cerebellum (page 480) and are passed by here.

The major part of the sensory tracts, which, *uncrossed*, run through the *posterior columns* of the spinal cord (paths for the *muscular sense*) and end in the *posterior column nuclei* (spino-bulbar tract) takes from these its continua-

tion in the central (bulbothalamic) sensory path, which, above the pyramidal decussation, also crosses as fibræ arcuatæ internæ (*superior pyramidal decussation*) and forms the *inter-olivary layer* (see Figs. 123 and 124). These fibres, after crossing reach the medial or *principal* fillet (Fig. 138), which ends in the *optic thalamus*. For the situation of the main fillet in the pons see Fig. 137.

That part of the sensory fibres that has crossed in the spinal cord (tract. spino-thalamicus), and passes through the lateral tract (pain and temperature conductors), consisting possibly of a chain of short tracts,

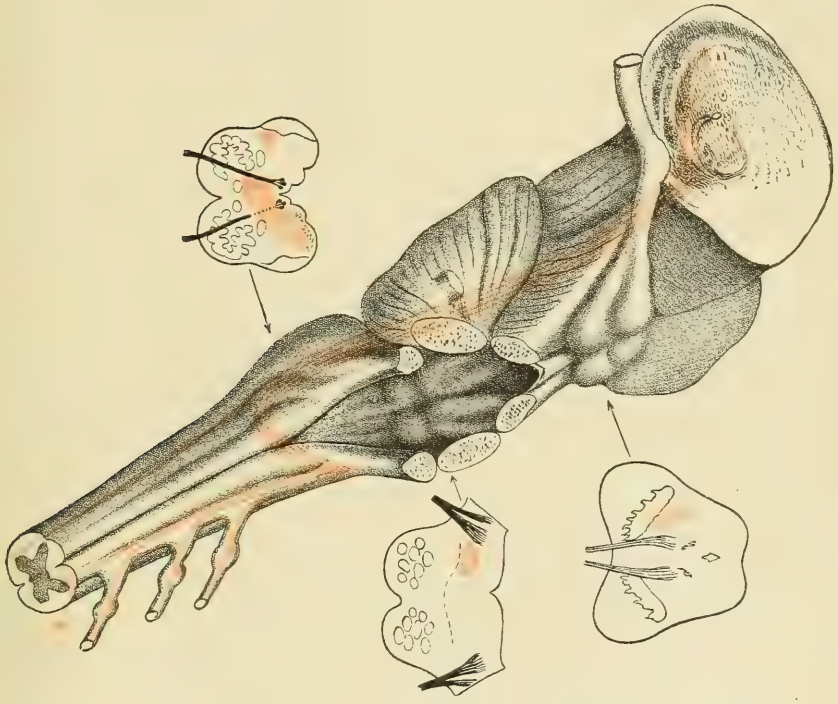


FIG. 138.—General course of the medial or principal fillet. (*After Edinger.*)

reaches the *reticular formation* of the pons and joins the principal fillet, in order to reach the *optic thalamus together* with it.

Touch sensations seem to pass through both, posterior column and lateral tract.

The separate position of the paths for the muscular sense on the one hand (inter-olivary layer, fillet), for pain and thermic sensations, on the other (reticular formation—subst. reticularis), in the medulla and the more caudal planes of the pons, makes it possible, that a focal injury here may cause such a dissociation of sensibility that position and movement sensations are retained, and pain and thermic sensations lost, or vice versa; since the paths for the muscular sense cross only in the medulla, it depends on the plane, in

which the focal injury lies, whether the sensory disturbance is on the crossed or uncrossed side. The fact that the paths of the lateral tract have already crossed in the spinal cord, makes possible very complicated results (as loss of muscular sense on the same side, and of the thermic and pain sensations on the opposite side).

Focal injuries in the pons, which strike the *trigeminus* roots or nuclei and the already crossed sensory paths, cause sensorial disturbances in the face, on the same side as the injury, and in the body, on the opposite side (*hemianæsthesia cruciata*) (Fig. 140).

The central path that rises in the *nuclei of the sensory cerebral nerves* (fifth nerve, etc.), and probably also, as has been said, on proximal planes of the pons, the central continuation of the lateral tract, that has reached the

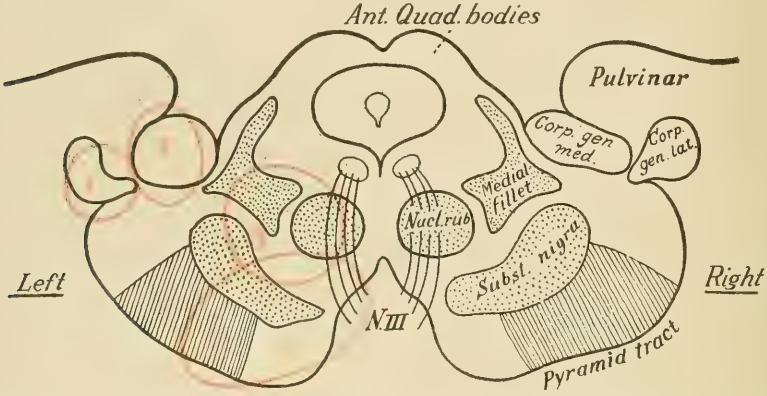


Fig. 139.—Foci in the quadrigeminal region. Focus 1 produces hemianopia (at the right side). Focus 2 diminishes the auditory acumen in both ears. If combined with a lesion of the auditory tracts at the right side it produces bilateral deafness. Focus 3 produces disturbance in sensibility at the right side (because of the lesion of the fillet) and paralysis of the oculomotor at the left side. Focus 4 produces disturbance of motility in the right arm and leg and lesion of the oculomotor at the left side (hemiplegia alternans superior).

reticular formation, and is there interrupted by gray matter, join the principal fillet. Thus all the sensory paths reach the *thalamus*.

On the entire way to the thalamus, the sensory paths are separated from the motor paths and can, therefore, obviously be interrupted by injuries that spare the motor paths. Thus, for instance in the frontal division, throughout the hindmost part of the thalamus, the sensory paths lie in a position clearly dorsal (in the tegmental region) from the pyramidal paths that go through the foot of the cerebral peduncle.

From the thalamus, the *thalamo-cortical* path passes through the posterior limb of the *inner* capsule, medially from the lenticular nucleus, and partly *through it* to the cortex.

The former supposition that the sensory paths in the posterior limb of the inner capsule are placed absolutely separate from the motor paths, and lie *behind* the latter (Fig. 134), has been impossible to maintain.

The isolated sensorial losses often noticed in focal injuries situated here (with motility intact) are caused, in the majority of cases by the participation of the optic thalamus in the focal injury. But even if the sensory paths of the inner capsule run partly together with the motor paths it is still possible for them to be struck comparatively isolated and en masse, in the place, where they pass from the ventral optic thalamus nucleus into the posterior limb of the inner capsule, so that focal injuries in the hind third of the *posterior* limb of the inner capsule cause more damage to sensibility than to motility.

Because of the proximity of the central visual path passing through the hindmost part of the inner capsule, the capsular sensory disturbances are generally combined with hemianopia.

The ends of the sensory paths, lie mainly and most densely in the posterior central convolution. The anterior one receives only few sensory fibres. But besides the posterior *central convolution*, the anterior division of the parietal lobe receives them.

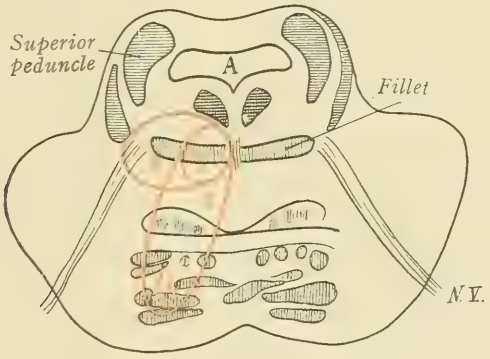


FIG. 140.—Effect of a focus in the middle of the pons. (Exit of the trigeminal nerve.) *A*, Aqueduct; *B*, pyramidal tract. Focus 1, striking only the fillet, produces crossed disturbance of sensibility in the body. Focus 2, striking besides the root of the trigeminal, produces disturbance insensibility of the face on the same side, crossed disturbance of sensibility in the body (Hemianæsthesia cruciata). Focus 3 produces crossed motor and sensory disturbance.

Focal injuries in the posterior central convolution, affect with special force, sensations of place, position and movement.

Localization by the patient, in that case, is extremely poor, and the consciousness of the position of the limb is often wholly lost. On the contrary, the other qualities, pain, temperature, and touch sensations, are attached to extensive cortical fields (both central convolutions) and other regions, especially of the upper parietal lobe, and are affected more or less slightly in mere focal injuries of the posterior central convolution. Great destruction of the posterior central convolution, besides that of the adjacent parts of the parietal lobe can cause serious hemianæsthesia, which, however, as has been said before, may retrogress to a considerable degree, in the course of time, except that the loss of muscular sense is more permanent. (Some

authorities hold that the anterior central conventions have no sensory functions.)

When sensations of place, position and movement are lost, and those of touch injured, naturally the forms of objects are not recognized, by feeling them, and the objects themselves can not be identified. This is a perceptive astereognosia which is a natural consequence of the loss of the elementary qualities needed for recognition of form, and must not be confused with Wernicke touch-crippling (touch-paralysis, *tactile agnosia*). It is to the latter what the blind man's inability to read is to alexia.

The above mentioned tactile agnosia, appearing in lesions of the middle third of the posterior central convolution and its immediate vicinity towards the rear, cortical touch-paralysis, is discussed hereafter.

Figs. 139 and 140 illustrate the effects of variously situated focal injuries in the quadrigeminal region and the pons.

(e) Cerebral and Cerebellar Ataxia

As destruction of the posterior column of the spinal cord, through the loss of centripetal signals, which regulate movement, especially of the stimuli coming from the muscles, joints, ligaments, causes spinal ataxia, so extensive breaks in these sensory paths, in the higher divisions—in the medulla, in the cap of the pons, in the cerebral peduncle, in the thalamus, in the inner capsule, centrum semiovale and cortex—cause cerebral ataxia. Hence, especially, lesions of the fillet, optic thalamus, thalamo-cortical tegmental path, the cortex of the posterior central convolution and of the adjacent parietal cortex, disturb the co-ordination of movements.

Cortical ataxia and subcortical ataxia, which is caused by a break in the sensory paths and is with difficulty differentiated from it present a somewhat different picture from that of the well known spinal ataxia of tabes. The excessive, flail-like movements of the tabetic are not so prominent. Instead, all movements become inexact, without consideration as to the shortest way; now the movement comes near the goal, now it falls short of it.

The fine harmony in the co-operation of muscles is absent; neither choice, nor measure, nor sequence in time of the innervations is correct. The synergies (co-operation of muscles (agonists) and their antagonists and of collateral and rotatory synergists) are disordered. Often the object falls from the hand, the sensory disturbance directly co-operating with ill managed muscular action. The writing of the ataxic is scribbling, shows evidences of trembling and does not stay on the line; the letters and parts of letters are uneven and contorted. All the assurance, exactness and finish, that practice has given our movements, are lost. In what was formerly called cortical ataxia, there was included, besides this awkwardness, which is conditioned by the loss of centripetal control, something, now classed with apraxia:

kinetic memories for certain innervation complexes are lost, cf. apraxia. (Probably mimic disturbances, at times observed in focal injuries of the thalamus are merely the consequence of loss in sensibility.)

Different from cortical and subcortical *cerebral ataxia* (focal injuries in the medulla, pons, etc., belong here, in as much, as they act through lesions which affect paths going to the cerebrum) is *cerebellar ataxia*. The cerebellum has less to do with the *co-ordination* of the movements of the extremities, than with the regulation of *upright position*, of the *maintenance of equilibrium* in *standing* and *walking*. The sufferer from cerebellar disease totters like a drunken man. The characteristic of cerebellar ataxia is, that a person exhibiting it in its severer forms when walking or standing, shows comparatively little ataxia in arm and leg movements made while lying on his back.

Especially in walking interference is shown in the *regular co-operation of legs and trunk*. This may be only unilateral and then the ataxia is on the same side as the cerebellar focal injury (in contradistinction to cerebral focal injuries). This control by the cerebellum occurs probably because of the stimulations flowing towards it through the lateral cerebellar tract, *which do not become conscious*, whereas the usual control of the movements of the extremities comes from the cerebrum and is based for the most part on *conscious* perceptions.

f. Symptoms of Lost Sensory Function (Disturbances of the Special Senses)

1. *Visual Disturbances, Visual Paths*

(a) **Choked disc** has been discussed under general symptoms. See page 487.

In long persisting choked disc, central keenness of vision is generally impaired, and the field of vision irregularly restricted. The final result may be blindness, which corresponds ophthalmoscopically to the clinical picture of atrophy of the optic nerve. If an operation be had in time or perhaps in case of cerebral syphilis, therapeutical methods be used, choked disc and optic neuritis may disappear entirely. [It should never be forgotten that a quite marked optic neuritis may be present and the patient not realize any deficiency of vision.]

(b) **Atrophy of the optic nerve,** greenish *whitish* discoloration of the well defined disc, usually bilateral, is a rather frequent symptom of degenerative processes in tabes and tabo-paralysis, occurring less often in pure syphilis. It often precedes the other symptoms by years. One must be very cautious, therefore, in assuming the existence of isolated optic atrophy without tabes, paralysis, or syphilis. It may happen, however, that optic atrophy exists permanently without other nervous disease. Optic atrophy of slighter degree is found, at times, in multiple sclerosis.

There are, besides, other bilateral diseases of the optic nerve, which cause slight or no changes in the fundus of the eye, that often lead to sudden blindness or produce only central scotoma especially for red and green.

They are traced back to certain neuritic processes in the optic nerve (neuritis retrobulbaris), toxic substances, as alcohol, nicotine, extract. filic. mas., being usually the cause. A more detailed description of them belongs in a text-book on diseases of the eye.

Of interest is "*amaurotic idiocy*," recently discovered, a combination of optic nerve atrophy with peculiar changes in the macula lutea and idiocy, which in certain families is hereditary and found chiefly in Jewish children.

(c) Injuries to the **chiasm.**

Since affections that attack the chiasm, generally proceed from the middle, very frequently from the hypophysis, it frequently happens that both crossed bundles of the optic nerves—which supply the nasal retinal surface—are affected, therefore the temporal halves of the field of vision are blinded (bitemporal hemianopia). If the uncrossed bundle on one side is also affected, total blindness of the eye on that side, and only temporal blindness of the other results. A lesion arising without the chiasm that affects it (arrow Fig. 141), may cause unilateral nasal hemianopia.

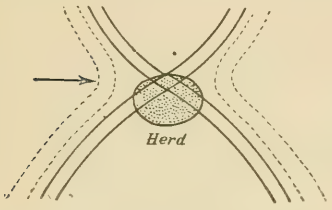


FIG. 141.—Lesion in the chiasm arising from disease in the hypophysis and producing bitemporal hemianopia. The uncrossed fibres are indicated by dotted lines. The crossed fibres affected by the lesion subserve both nasal halves of the retina, therefore the temporal halves of the visual field.

The combination of bitemporal hemianopia with acromegaly is well known as characteristic of a tumor of the hypophysis.

The Visual Paths.—Into the optic tract of each side, enter the fibres of the same sided half of the retina of that side and the (crossed) fibres of the same sided half of the retina of the opposite side. As to their main body, the tractus fibres end in the *lateral geniculate body;* a smaller part ends in the *anterior quadrigeminal body and in the pulvinar.* See Fig. 142. They are the so-called *primary optical centers.* The fibres, that go to the quadrigeminal body, come into communication with the oculomotor nucleus, and produce the pupillary reflex.

From these primary centers the so-called optic radiation passes back into the occipital lobe, and, surrounds, in its entire course from the outside, the posterior horn. In this course it passes the deep white matter of the lower parietal lobe (especially of the angular gyrus) and occipital lobe and the greater part of it reaches the medial surface of the occipital lobe, in the vicinity of the *calcarine* fissure, in the visual center marked by Vicq d'Azyr's stripe and peculiar formation. This embraces mainly the *cuneus* (dorsal lip of the calcarina) and the *lobus lingualis*, perhaps also the fusiform gyrus. The

visual center encroaches but little upon the convexity, except perhaps at the occipital pole and in the third occipital convolution.

From the visual center pass *corticofugal* paths, together with the cen-

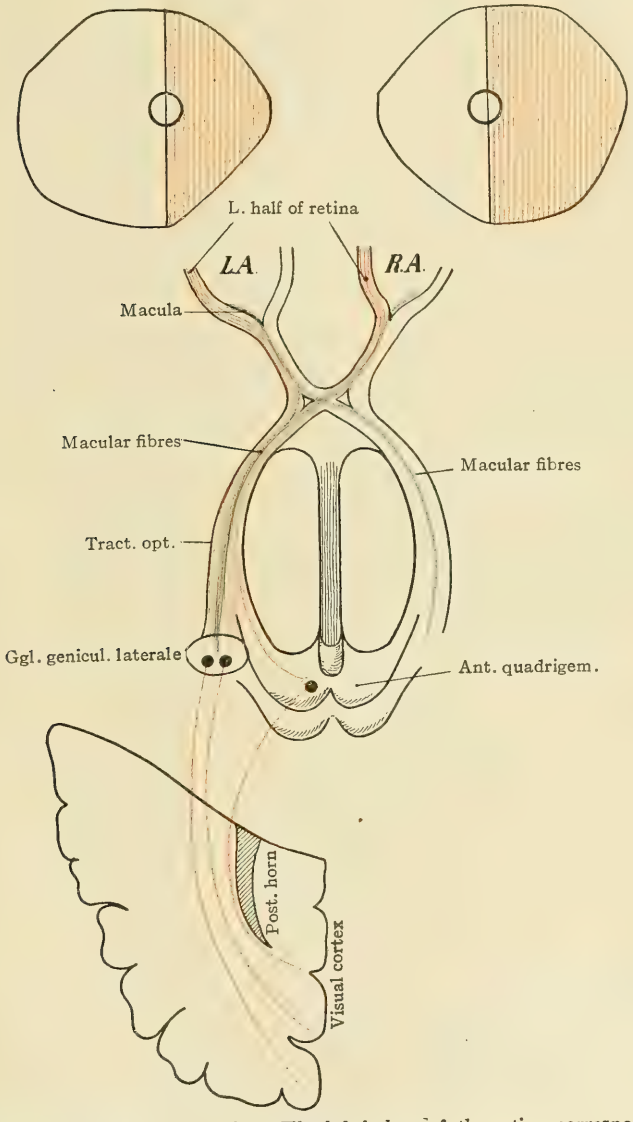


FIG. 142.—Diagram of the optic paths. The left halves of the retina correspond with the right halves of the visual field.

tripetal ones, just described, into the same ganglia, from which the optic radiation arises.

The posterior horn is surrounded, on the outside, by three *medullary layers*, the fibres of which run *sagitally*. The innermost, lying directly

adjacent to the ependyma of the ventricles, lining the ventricles to a certain extent, is called the tapetum (Strat. sagittale mediale).

Towards the outside of it, therefore in the middle, lies the *stratum sagittale internum* and outside of it, the *stratum sagittale externum* or *inferior longitudinal fasciculus* (Fig. 143).

The latter was long considered a pure bundle of association fibres between occipital and temporal lobes, whereas the real optic radiation (Gratiolet's) was placed in the stratum sagittale internum. The tapetum was supposed to consist of corpus callosum fibres exclusively. But it has been proved, that in all three strata, especially in the stratum internum and externum, optic radiations course, that the stratum sagittale externum also contains association fibres, the tapetum, mostly corpus callosum fibres, by intervention of the forceps. At any rate, the optic radiation can no longer be

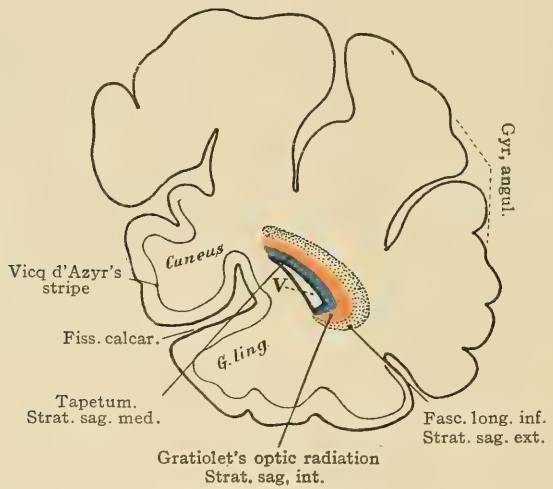


FIG. 143.—Diagram of the three sagittal medullary layers surrounding the posterior horn *V* in a frontal cut through the gyrangularis.

confined to the middle layer, but to the inferior longitudinal fasciculus must be assigned a large part of the optic projection fibres.

Lesions *behind* the chiasm throughout the visual path, that is in the optic tract, in the ganglion geniculatum laterale, in the optic radiation and the cortical center, are bound to cause *bilateral homonymous-hemianopia*, that is loss of function in the halves of both retinas, on the same side as the cerebral lesion and correspondingly, in the halves of the field of vision on the opposite side. In the majority of cases, the whole half of the field (except the so-called superfluous field) is lost, so that hemianopia, in the strict sense, occurs. That is, when the focal injury is on the left, a right hemianopia of both eyes corresponding to the blindness of both left halves of the retinas.

Under certain conditions, only a quarter is lost for both eyes: *quarter hemianopia* (Fig. 144).

It is important to notice, that the loss of *exactly* half of the field of vision

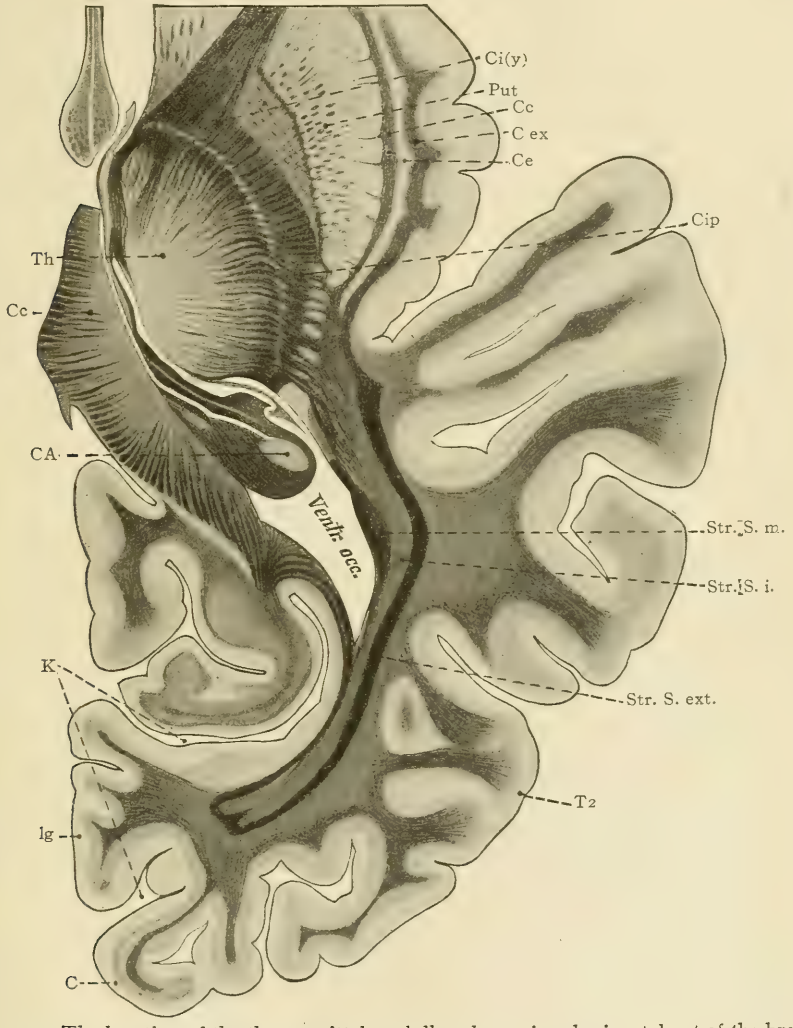


FIG. 144.—The location of the three sagittal medullary layers in a horizontal cut of the brain (Drawn from a Weigert preparation (3/2 of natural size) *from Déjérine.*) *C*, cuneus. *CA*, Ammon's horn. *Cc*, corpus callosum. *Ce*, external capsule. *Cex*, capsula extrema. *Ci(y)*, capsula interna (genu). *Cip*, capsula interna (posterior limb). *Cl*, claustrum. *K*, calcarine fissure (appears in two places). *Lg*, gyr. lingualis. *Put*, putamen (external layer of the lenticular nucleus). *O*, occipital convolution. *St. s. ext*, stratum sagittale externum or fasciculus longitudinalis inferior. *Str. s. i.*, stratum sagittale internum or Gratiolet's visual radiation. *Dtr. s. m.*, Stratum sagittale mediale or tapetum. *T2*, second temporal convolution. *Th*, thalamus.

(total hemianopia) is very rare. This would mean, that the line of division between the parts of the visual field that are retained and those that are lost, would go as a vertical line directly through the point of fixation. It occurs

occasionally, with focal injuries in the optic tract, but not generally with centrally situated focal injuries. Usually at least a small area about the point of fixation, in the direction of the loss, belongs to the *part retained.* This very area, namely, the part retained, situated beyond the vertical

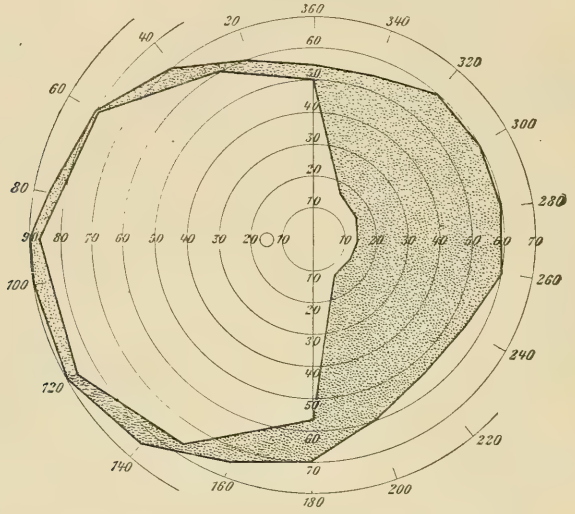


FIG. 145.

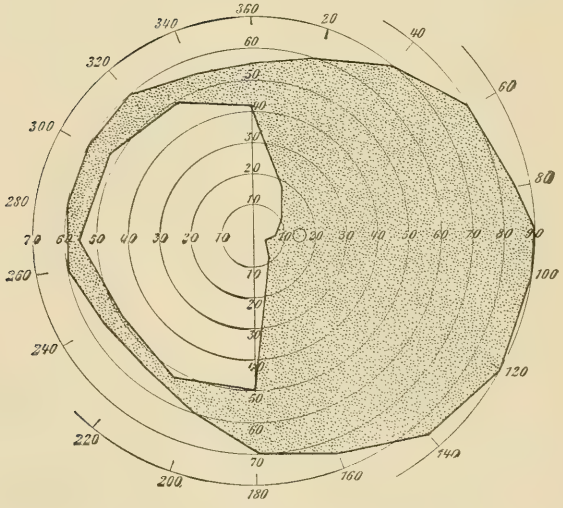


FIG. 146.

FIGS. 145 and 146.—Visual field in right-sided hemianopia with residual field of vision.

meridian passing through the point of fixation, is known as the "superfluous field of vision." The line of division makes a curve about the point of fixation, at least, which with its convexity extends into the blind half of the field of vision (Figs. 145 and 146).

The fact, that the direct vicinity of the point of fixation on both sides, is almost always retained, proves that the region of *clearest sight* in the retina (macula) is *double*, that is, represented in both hemispheres. The size of the superfluous field of vision varies with the individual case (2 to 15 degrees) and may even be different in each eye. Moderate concentric narrowing of the retained field of vision, also may occur, more obviously in the eye that is on the opposite side from the focal injury. Certain other irregularities in the boundaries of the visual field, in focal injuries of the occipital lobe, are still more difficult to understand.

Bilateral hemianopia is called *cortical blindness*, an adopted, but not wholly correct nomenclature (cerebral blindness is better), since it is often a question, not of cortical focal injuries, but of bilateral interruption of the optic radiation or cortical focal injury on one side and *sub*cortical on the other.

In this bilateral hemianopia a small central part of the field of vision (sometimes 2 to 3 degrees) is generally retained, which, however, easily escapes both observer and patient, and, therefore, demands skilful investigation to prove its existence.

In rare cases, both halves of the visual field have been lost *simultaneously*, by embolism of the occipital lobes.

Vision in the retained visual field is often, though not always, diminished in hemianopia as well as in cortical blindness.

A case in which, on the contrary, macular vision is destroyed, but peripheral retained—that is a field of vision with central scotoma—due to an occipital focal injury—has not yet been observed to occur as a *permanent affection*.

The fact that in uniocular as well as in binocular hemianopia, macular vision is usually retained, has been attributed by some investigators to double cortical representation of the macula, by others to the fact, that the situation of the cortical representation of the macula, is protected especially against circulatory disturbances, by others (who speak against an isolated representation of the macula), to a representation of the macula, extending over large areas of both occipital lobes.

From the last point of view, the macula has an extraordinarily extended representation in the occipital lobe, so that only immensely extended focal injuries could rob it of all connections with the cortex.

How far definite parts of the retina correspond to definite parts of the cortical visual center, i. e., how far the retina is, strictly speaking, projected on the cortex, has not been unanimously decided. There is much difference of opinion about it.

As the termination of the optic radiation (visual sphere, visual center) chiefly the vicinity of the calcarine fissure must be considered. Especially its upper border—the cuneus—and its under border, the fusiform gyrus,

probably the entire region marked out by the Vicq d'Azyr stripe, the cellular structure of which is peculiar and uniform.

The fact that quarters and even sixths are lost by focal injuries in the optic radiation and visual center proves, at any rate, that no complete mingling of fibres, which come from various parts of the retina, takes place, that at least, on broad lines, projection takes place. And so to the upper quarters of the retina in both eyes, corresponds the upper border of the calcarine fissure, to the lower quarters, the lower border.

By important authorities, the pathological discoveries which led to this proof, were used to show that not the cortical focal injuries concerned, by themselves, but accompanying lesions of various parts of the *optic radiation*, had the effect mentioned, so that a projection of the quarters would exist in the optic radiation, but no longer in the cortex.

The last word has not yet been said about the projection of the macula.

Instead of hemianopia, mere *disturbances of color vision* may affect half the field of vision (hemiachromatopsia or hemidyschromatopsia). Now general weakness for color vision appears, now color blindness for red and green only, or less frequently for yellow and blue. It is remarkable that often with right hemianopia, is found hemidyschromatopsia in the half of the field of vision that has been retained.

Paralyses of the muscles of the eye are *not*, per se, connected with hemianopia. But the loss of sensory-optic stimulations can produce an uneconomic use of the muscles of the eye and hence lead to mistakes in estimating distance.

With hemianopia, from whatever cause it arises, frequently *disturbance in measuring with the eye* is connected, in such a way that by halving a horizontal, the half corresponding to the loss in the field of vision is made too small. (The opposite error is rare.)

Hemianopia caused by a focal injury in the optic tract must be distinguished from hemianopia caused by an interruption in the optic radiation or injury in the visual center, by the fact that in the former case, throwing light upon the blind half of the retina, causes no pupillary reflex, whereas there is no cause for this in the latter case, since the reflex arc (connection of the tract with the oculomotor nerve) is intact. The proving of pupillary rigidity in hemianopia is, however, attended with great difficulties, because it is impossible to succeed in confining the illumination exclusively to the retinal halves concerned. But in bilateral hemianopia, caused by cortical or subcortical focal injury, the fact that the pupillary reflex is retained, can be easily proved.

Subcortical focal injuries may *irritate* the visual center, and occasionally cause visual hallucinations in those parts of the field of vision that have been lost.

It is still uncertain which part of the visual system is affected in uræmic amaurosis.

In addition to the hemianopsia present in all focal diseases, which strike the visual system from the tractus to the visual center, it is found frequently as a transient affliction in progressive paralysis, after the *attacks*, and in migraine. After focal injuries in the temporal and parietal lobes, it occurs temporarily in the first weeks after the attack as an indirect symptom of the nearby lesion. Hemianopia is frequently the first symptom of apoplexy. Focal lesions in the parietal lobe, especially in the angular gyrus, which extend *deeply below the surface*, break through the optic radiation, and hence cause *permanent* hemianopia (Fig. 147).

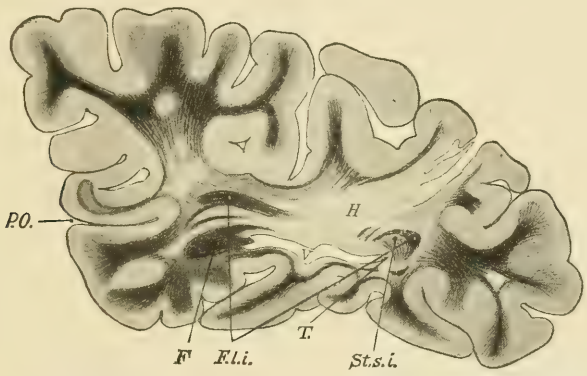


Fig. 147.—Case of softening in the gyr. angularis. The softening focus *H* passes through the 3 sagittal medullary layers. Drawing from a Pal preparation. Nat. size (frontal cut). At the left, medial surface of the brain. *V*, posterior horn. *P.O.*, fissura parieto-occipitalis. The fissure without any designation below the latter is the calcarine. *F*, forceps, the preserved part of the corpus callosum radiation. *F.l.i.*, The preserved parts of the fasciculus longitudinalis inferior. *Str.s.i.*, Remains of the stratum sagittale interius. *T*, place of the absent tapetum.

2. *Auditory Disturbances*

Auditory Paths.—The cochlear nerve (which is the only branch of the acoustic nerve that is concerned with hearing), rises in the cells of the spiral ganglion of Corti. It ends in the *ventral* nucleus of the cochlear nerve and in the *dorsal* nucleus (acoustic tubercle-tuberculum acusticum). From these spring other paths, which, excluding further interruptions in the nuclei of the lateral fillet, for the most part pass in the trapezoid body (interruption in the olive), partly pass into the striæ acusticæ, on the other side, reach there the opposite *lateral fillet*, and for the most part end, after going through the arm of the posterior quadrigeminal body, in the *medial geniculate ganglion* (the smaller part in the posterior quadrigeminal body) (Fig. 148).

Part of the fibres remain *uncrossed*, and there is, besides, a commissure between the bilateral nuclei of the lateral fillet, so that each cochlear nerve *is connected with* both medial geniculate bodies, hence with both *hemispheres*. Here, too, we find *semi*crossing.

From the medial geniculate ganglion the central auditory path goes to the cortex of the temporal lobe and the greater number of the auditory fibres reach the *transverse convolutions* of the temporal lobe (gyr. transversi), Heschl's convolutions, and a neighboring tract in the middle third of the first temporal convolution.

The further course of the fibres reaching the posterior quadrigeminal body is a matter that has not as yet been cleared up.

Deafness in the opposite ear, after *one-sided* lesions in the temporal lobe, has never been observed. In that case, we have only lessened hearing, in both ears, corresponding to the connection of each ear with both temporal lobes.

Likewise, unilateral interruption of the medial geniculate ganglionic paths, going to the temporal lobe, does not cause total one-sided deafness.

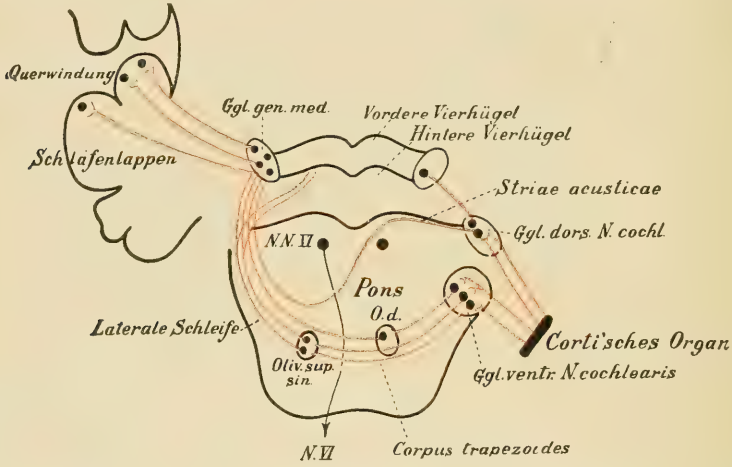


FIG. 148.—Scheme of the central auditory paths. Interruption in the nuclei of the lateral fillet not considered. *N.N.VI.*, nucleus of the abducens. *O.d.*, right superior olive. In the cut: Laterale Schleife = lateral fillet. Schläfenlappen = temporal lobe. Querwindung = transverse convolution. Vordere Vierhügel = anterior quadrigeminal bodies. Hintere Vierhugel = posterior quadrigeminal bodies. Cortisches Organ = Corti's organ.

Total deafness in both ears may be caused by bilateral disease in the temporal lobes (*so-called cortical deafness*). Heretofore, this has been observed only in very extended focal injuries, so that the above anatomical boundaries of the auditory center (based upon the myelogenetic method) could not be clinico-pathologically substantiated, and many authors ascribe to the auditory center much more space in the temporal lobe, than in following *Flechsig*, is given above.

More often than total deafness, great difficulty in hearing, in both ears, results from bilateral lesions of the temporal lobes. This does not show the well known tone-gaps of labyrinthine diseases (as the loss of the highest tones), but a more regular diminution for the entire tonal sequences.

This does not decide against a projection of various tone-heights on the cortex, since in the two-sidedness of the representation of the organs of Corti

in *both* temporal lobes, an insulated tone-gap could only appear, if by chance, exactly the same parts were destroyed on both sides. At any rate the assumption, that is to be discussed under sensory aphasia—that the cortical end-

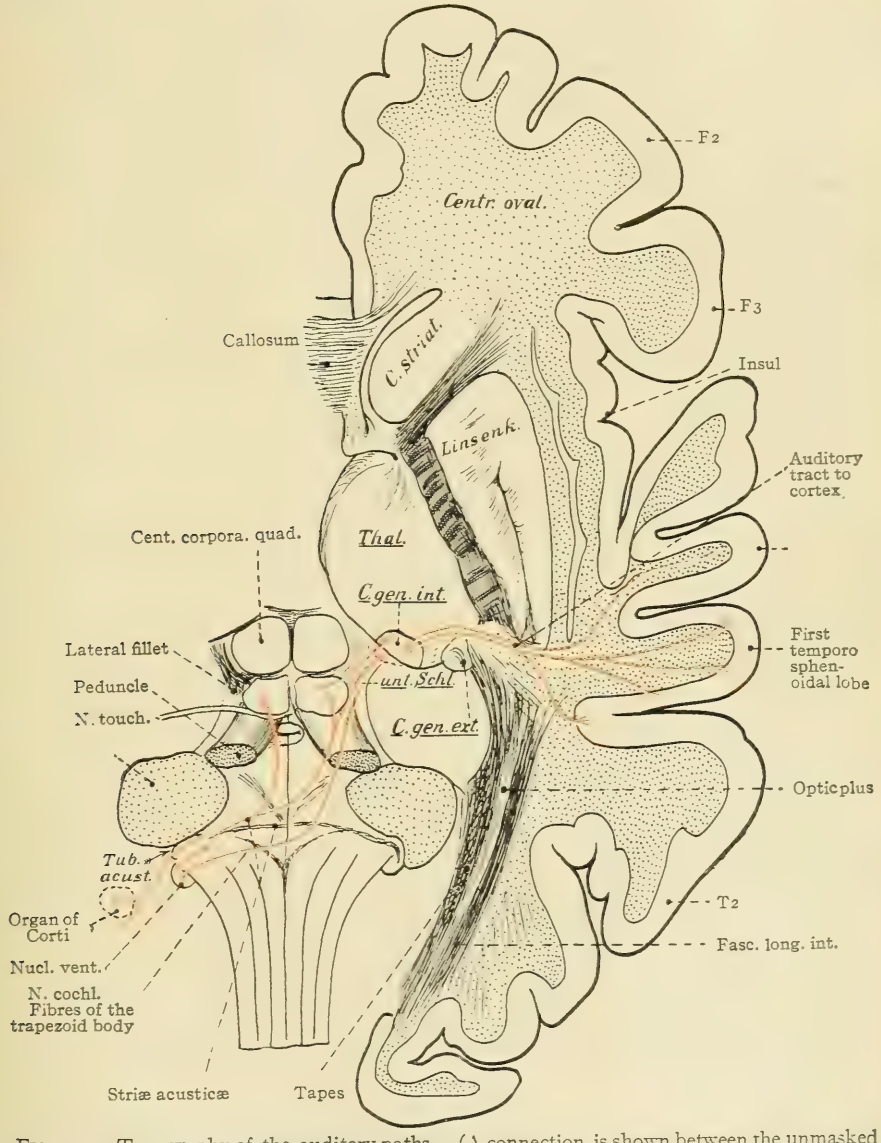


FIG. 149.—Topography of the auditory paths. (A connection is shown between the unmasked posterior quadrigeminal body and the *C.* gen. int.) In the cut: Linsenk. = lenticular nucleus. unt. Schl. = lower fillet.

ing of the fibres of the tones b′ to g″—a very important part of the tonal sequence—take up a definite spacial division within the entire auditory sphere, is as yet, wholly hypothetical.

Auditory disturbances caused by focal injuries in the cap of the cerebral

peduncle and the pons and in the region of the posterior quadrigeminal body, occur, yet, because of the wide distribution of the paths concerned, and their semi-crossing, are seen permanently in high degree, only when the focal injury is very extensive—except when nucleus or *root of the cochlear nerve* is destroyed, in which case, deafness in one ear results.

3. *Olfactory and Gustatory Disturbances*

In basal focal injuries that affect the olfactory nerve, nucleus, or tract, the olfactory trigone, or the perforated substance, a diminution in the sense of smell, on both sides, is noticed.

The olfactory centers lie in the hippocampal gyrus and the Ammon's horn.

Olfactory hallucinations also appear in irritations of the olfactory regions.

The gustatory sphere, too, lies in Ammon's horn, the hippocampal gyrus and the adjacent posterior end of the fornicatus gyrus. The peripheral paths of taste course in the trigeminus and the glosso-pharyngeal nerves.

3. Mnemic-associative Derangements

(a) Aphasic Disturbances

1. *Introduction*

Disorders of the Projection System of the Apparatus of Speech: Anarthria in Bulbar and Pseudo-bulbar Paralysis

Language is a system of symbols serving for the communication of thought between men.

The symbols of spoken language are sounds and combinations of sounds which are naturally produced by the action of the muscles of the lips, jaws, tongue, palate, and larynx. In this aspect, speech is, then, a series of movements, the effect of which is to give other people symbols of the psychic processes, such as thoughts, feelings, desires, etc., of the person speaking. (*Speech-expression.*)

But considered with reference to its effect as audible sound, it may be included with the process of hearing, and evokes in the listener the corresponding psychic processes. The rôle of the listener is understanding. (*Speech-perception.*)

Not until late in the development of both the species and the individual, do we find a second system of symbols: *written* speech which also has an expressive (writing) and a receptive (reading) side.

Both *written* and *spoken* language employ certain muscles which serve *other* purposes as well. Thus the muscles of the tongue, jaws, and lips, serve for eating, chewing, sucking, swallowing, licking, kissing, etc., and for various voluntary and involuntary imitative movements.

If this system of muscles or its corresponding nervous centers is diseased (paralysis, paresis, ataxia, tremor, etc.), speech is affected secondarily. The resulting disturbances of speech are called, according to their degree, *dys-* or *an-arthria*.

To this nervous mechanism belong:

1. The cortical centers of the muscles concerned, therefore the centers of the 12th, 7th, 10th, and 11th Nerves in the lower fourth of the anterior central convolution. (Operculum Rolandi.)

2. The motor (cortico-bulbar) tract from these centers to the bulbar nuclei of the nerves. They pass through the white matter, through the "genu" of the internal capsule, the base of the cerebral peduncle and reach to the nuclei of both sides in the pons Varolii and the medulla oblongata.

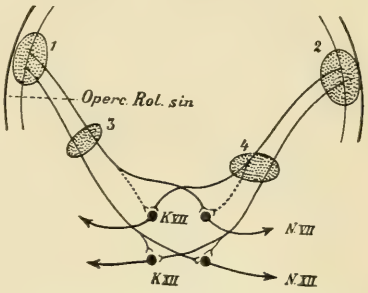


FIG. 150.—Seats of lesions causing pseudo-bulbar paralysis. 1 and 2 foci in the left and right cortical center of the seventh, twelfth nerves, etc. 3 and 4 foci in the paths to the pontine and bulbar nuclei. The combinations: 1 + 2, 2 + 3, 1 + 4, 3 + 4 produce pseudo-bulbar paralysis.

Because of the connection of each hemisphere of the brain with the nuclei of both sides, it results that supranuclear lesions of one side do not produce permanent paralysis of the nerves concerned, and therefore the dysarthria resulting is neither severe nor long-continued. A break in the connection of the cortico-bulbar tracts and their centers can indeed bring about temporary dysarthria, especially when the left hemisphere is affected. Slight dysarthric disturbances often persist after a lesion of one side.

3. The peripheral tract from the nucleus of the 12th Nerve, etc., to the muscle.

Disease in the medulla oblongata affects the peripheral tract represented by 3. If the bulbar nuclei, or their roots, are destroyed, we have *bulbar paralysis*, together with atrophy and degenerative reactions in the muscles concerned (tongue, lips). It causes besides the *dysarthria*—which consists of incorrect articulation of letters, and is conditioned by the centers affected, whether they be more particularly labial, palatal, or lingual—*other paralytic phenomena:* partial paralysis of the facial muscles and disturbances in eating, chewing and swallowing.

If the cortical centers or the supranuclear nerve tract are affected *on one side alone*, there results only, as we have said, a slight degree of disturbances in the muscles concerned: slight paralysis of the ends of the facial nerve, deviation of the tongue, when it is extended, slight difficulty in articulation with no disturbance in swallowing, no atrophy and no reaction of degeneration. But if the centers or nerve tracts are disturbed *on both sides*, or on one side the center and on the other the tract, similar disturbances result as arise

from the destruction of the bulbar nuclei themselves, only that atrophy and degenerative reaction are absent and that there is a smaller degree of disturbance. Since the supranuclear fibres above the pons disperse in all directions, very great focal injury is necessary to interrupt them *wholly*. Those localized in a supranuclear position in the pons are most likely to effect this (Fig. 150).

Since such two-sided disturbances of the cortical centers and the cortico-bulbar nerve tracts produce phenomena so like bulbar paralysis, they have been named *pseudo-bulbar paralyses*.

The essential feature of pseudo-bulbar paralysis is therefore that *on both sides* the cortico-bulbar nerve tracts (eventually at the place of their cortical origin) are interrupted at some one place. To bring this about, unless it be in the pons the nerve tracts must be stricken twice, once on the left, and once on the right side. In this case, there are disturbances in articulation, in swallowing, etc.

Disturbances in this efferent neuro-muscular apparatus belong to the derangement of the *projection system* alone and have no connection with aphasia, even when they are caused by cortical injuries. They cause cortical, cortico-bulbar and bulbar *anarthria or dysarthria*.

Not only spoken but written language as well employs a neuro-muscular apparatus, which serves other purposes as well. It is the apparatus which innervates the more delicate movements of the hand. Paralysis, weakness, ataxia of the hand, produce the same hindrances in writing, that pseudo-bulbar or bulbar paralysis produces in uttered speech, except that only rarely is the other hand affected also, and that the movements used in writing can be performed, more or less, by any other movable portion of the body (foot or tongue), while the production of sound is confined to a definite muscular system. Just as utterance can be affected indirectly by injury to the efferent neuro-muscular apparatus, the perceptive faculties of speech suffer from lesions of the *perceptive* apparatus, i. e., of the organs of hearing and seeing. By a difficulty in hearing verging on deafness, or as we shall see, by a definite *gap in the perception of tones*, the understanding of speech can be destroyed, as well as the ability to read by a weakness of eyes, verging on blindness.

Even then, we cannot speak of aphasic, but of perceptive disturbances.

2. *Disturbances of the Mnemic-associative Speech Apparatus: Aphasia. Internal Speech*

When, however, the cortico-bulbar apparatus, on which sound-formation depends, fulfills all its functions, and yet the patient can only speak badly or not at all, *aphasia* exists.[1]

[1] Mutism from hysteria and mental disorders is not taken into consideration here.

What has failed, is so-called *internal speech*, i. e., something that has been learned, *a possession of memory*.[1]

We use internal speech to distinguish it from external speech. The latter is the speech-musculature's creation, which produced by the one, the other hears. The primary condition for external speech is internal speech, but loss of internal speech entails the loss of external speech.

Internal speech is ever present while we are thinking. Most of our thoughts present themselves in the form of speech. In the case of vivid thought impulses discharge themselves into the neuro-muscular apparatus, so that we have abortive innervations, which may rise into involuntary utterance of thought.

The internal word consists originally of two parts, closely connected:

1. Memory of the word-*sound*, or the *acoustic* word (a), the residuum of remembered words, that the child hears from his environment. It is the oldest word-possession and *the great majority of words are acoustically acquired.*

Moreover the meaning of the word, i. e., the corresponding concept, attaches itself to the acoustic word—as the word Bow-wow to the concept of the dog—so that for many words, before the child itself can speak, there arises a close connection between the concept (c) and the acoustic word. C—a is the first speech-association.

2. The memory of word-*movements* or the motor-word (m). The memory of the process necessary for the utterance of the word.

Usually this process is called *kinæsthetic*. It comprehends memory of perceptions reaching all the parts moved by us in speech, lips, tongue, palate, etc. The utterance of the word "parrot" gives us other sensations of place, movement, and contact than, say, the word "weasel."

In truth, we find in *consciousness* nothing besides memories of this perception complex. But beneath the threshold of consciousness there comes a richer, a more *material* memory possession. The frequently repeated sequence of innervations, which correspond with a definite sound-complex, leaves probably a lasting connection between the co-working innervational elements, so that the motor-word[2] is composed of kinæsthetic and innervational memories, of which only the former have a psychic equivalent.

The motor-word is greatly dependent on the acoustic word. The mouth cannot produce what the ear has not heard. The child learns to repeat a series of words he has heard. By this means a very close connection is produced between a and m (a—m). This is the word as it exists in the child.

[1] We have learned articulation, too. But it is not what we have learned that is lost in dysarthria, but the natural instrument, with which what has been learned is produced.

[2] This innervational memory holds good in part for whole words, especially however, for parts of words, for syllables and letters.

This combination of the acoustic with the motor-word represents *internal speech* in its restricted sense. As we see, the concepts are predominantly connected with a, and with m only through the mediation of a, so that the relations of the motor-word to the concept is brought about mainly through a.

The following association is thus formed:

The recollection of word-movements can combine itself with the concept only secondarily, inasmuch as every concept has, indeed, led to m only through the mediation of a, yet the frequent simultaneous appearance of C and m nevertheless produces a secondary direct association. This less firm and comprehensive connection we shall represent by a dotted line between C and m.

When, between the ages of seven and eight years, the child learns the written language, by the complex (a—m) the *written* word attaches itself to the *sounded* word. But, unlike *hieroglyphics*, the written word-symbols are not direct symbols for the apprehension of the objects, but are *symbols for sounds*, i .e., for parts of the sounded word. In other terms, our writing is phonetic.

Now only does the child learn to separate the words into syllables and letters and to assign to each letter a written symbol. *Hence it is, that as a rule, the written language is connected with the concept only through the mediation of the sounded language.* To each sound an optical letter symbol is assigned (o) and in writing this optical letter is produced by the movement of whatever member is guiding the pen. Thus the acoustic motor-word, aroused by the concept, is separated into its component parts, each of which evokes its corresponding optical image, from which impulses flow to the hand center.

Through practice, *graphic-motor recollections* are formed in the hand-center.

The complex associations formed thus between the concept and the acoustic, linguo-motor, optical and grapho-motor components, represents *internal* speech in its *wider* sense.

Now impulses flow from m to the neuro-muscular apparatus of the tongue muscles. The stimulations in the hand-center flow off to the muscles of the hand, while to a and o arrive the centripetal stimulations of the ear and eye. The sound of the words penetrates to the seat of the acoustic memory images, arouses them, and thereby brings about the release of the components associatively connected with a, that is of the whole word.

The theory once held of a *direct speech pathway* to and from the bulbar nuclei had to be abandoned. On the contrary speech makes use of the same projection pathways (although with a certain preference for those on the left side) as the other movements of the tongue, lips, etc. Therefore the stimulation from m must first reach the cortical center "μ" of the 7th and 12th Nerves, etc., in the Rolandic Operculum, where the real innervation takes place. It is consequently necessary to insert "μ" between m and tongue.

In the same way the acoustic stimulations reach first the acoustic *projection-field* "alpha" before waking the remembrances in a. Even the authors who do not share the view of Flechsig of the absolute separation of the projection-field (acoustic-center) from the mnemic field (a), assign the perceptive and the mnemic process to various processes of the same field, so that perception and waking of the memory are regarded as *two* distinct

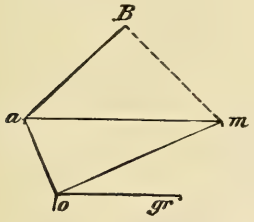


FIG. 151.—*B*, conception; *a*, acoustic component; *m*, speech motor component; *o*, optic component; *gr*, graphomotor component.

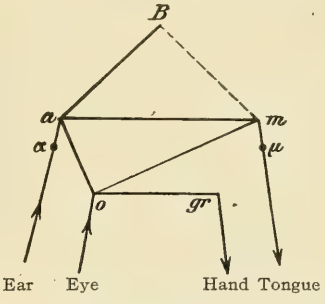


FIG. 152—*a*, acoustic perception; *u*, innervation. The other letters as in scheme Fig. 151.

parts of the process. (For o and gr the corresponding projection apparatus are not shown separately.)

The complete scheme of speech is shown in Fig. 152.

This scheme which is not intended to represent anatomic relations, but to make evident and comprehensible certain fundamental rules, plainly teaches the following: the way by speaking and writing leads from C[1] principally over a.

Speaking:—:B—a—m—"μ"—tongue

Writing:—:B— $\left(\begin{matrix}\text{a}\\|\\\text{m}\end{matrix}\right)$ >o—gr—hand

In the same way not only in understanding speech (Ear—"alpha"—a—B), but also in reading (Eye—o—a—B) the way leads over a.

m

Hence it is clear that an annihilation of a disturbs not only speech but

[1]B = Concept is represented in the above scheme only by a point, but in the brain, very extensive cortical areas correspond to it.

also reading and writing. But, since in writing and reading a dismembering of the entire word-sound (a—m) into its component parts (syllables and letters) takes place, also the loss of m will be peculiarly damaging for reading and writing. Writing and reading depend upon the condition that a and m are intact!

These relations of dependence (especially that B be directly connected only with a, further, that o enter into connection with B only over a—m) are not fixed *inevitably* by the organization of the brain, but by the way the function of speech is learned.

The deaf-mutes are the best illustration of this as they learn to write and even to speak, though a does not exist in them at all.

The relations of dependence can therefore be modified by individual predisposition, or *peculiar development.* For a time, it was believed even that individual peculiarities, sometimes the visual, then the acoustic, and again the motor predisposition gave grounds for questioning the regularity of speech mechanism altogether, that a great number of men with motor predispositions (Moteurs) could suffer the loss of acoustic speech components without injury to their speaking, so that they found innervations to speech directly from conception (i. e., in other words: the Concept evokes the motor components). Or that a great number of optically predisposed men (Visuels) are not disturbed in reading when a is injured, since they would be in a condition to evoke directly from conception visual images of letters. But it has been proved that the great *fundamental relations*, in consequence of similar formation in the majority of men, correspond more closely, than the exaggerated observations of individual differences have caused some to think. One can not even count four approximately similar types. There are only a few exceptions to the fundamental relations given in the scheme. The predominating place of a is almost always preserved and not overbalanced by individual predisposition, so that the motor word in the majority of cases, is conditioned by the connection between a and B, and written language usually also depends upon a. On the other hand, the rule that written language depends also on m, has not a few exceptions. There are people who suffer the loss of motor memories, without their written language being affected.

For the quicker or slower restoration, for the finer differentiation, and for the degree of the disturbances, individual differences must be considered. So must the *fluency in writing*, which a man possessed before illness, be considered also. As a rule, however, we may assume an approximate similarity in the organization of different human brains.

3. *The Speech Regions in the Brain*

Let us see, now, how the psychological scheme is carried out in the brain. Here, before all, a fundamental fact is to be noted: that the speech-functions

are performed chiefly by the *left* hemisphere. The opposite holds good only in left-handed people, in whom the right hemisphere plays the greater part.

Besides the left-handed people, 4 to 6% are ambidextrous—using both hands equally well—and only in these do both hemispheres share equally in the functions of speech. Now, it must not be thought that in right-handed people—the majority of mankind—the right hemisphere not only takes no part whatsoever in speech, but is also by nature unfitted for the function of speech. This is not true.

The right hemisphere, may, in healthy people, share in speech to a certain extent; but it is not able, by itself, to care for the functions of speech, so that from disturbance in certain parts of the left hemisphere, speech in

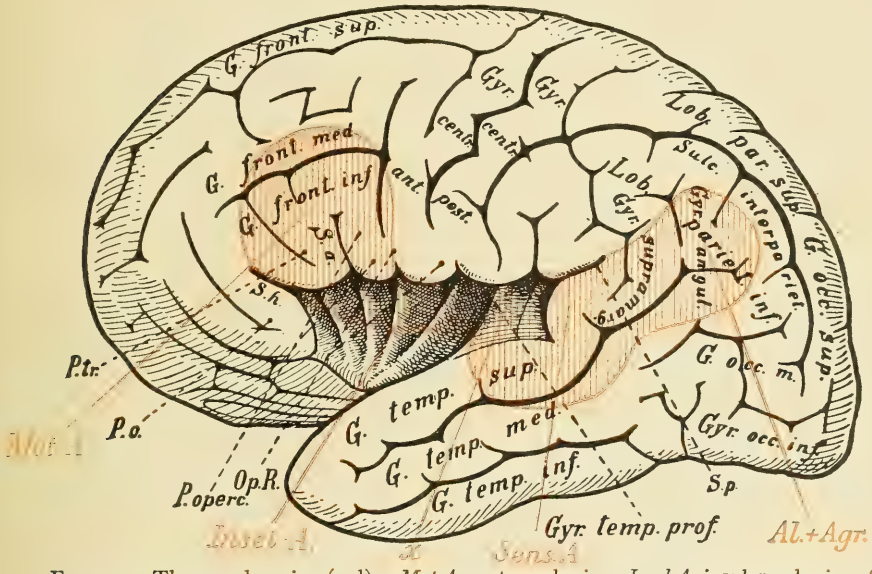


FIG. 153.—The speech-region (red). *Mot.A*, motor aphasia. *Insel.A*, insular aphasia. *Sens. A*, sensory aphasia. *A* focus in *X* (in the temporal transverse convolution) may perhaps cause pure word deafness. *Al. Agr.*, Alexia and agraphia.

certain of its functions (expressive, perceptive), is destroyed or badly injured. Were the right hemisphere as important to speech as the left, the same results would follow a lesion in the right hemisphere, which is not the case. Besides participating to a certain degree in the functions of speech in the healthy individual, the right hemisphere may, after a lesion in the left, undertake to some extent, after practice, the function of the injured half. There is, however, never a complete assumption in the case of right-handed people, and defects can always be proved by delicate tests.

All the rules to be stated hereafter hold good with only these limitations:

1. That ambidextrous people are not included.

2. That the right hemisphere (the left in the left handed) in the course of time, can to a certain extent, to be discussed later, substitute for the left.

3. That no individual differences in predisposition or use exist.

In the left hemisphere there is an area that is principally concerned with the functions of speech: the *speech region*, to which are to be added, those regions in which lesions *under* the cortex result in serious losses of function. But since the most varied combination of breaks in the fibres, so numerous as not to be discussed in a short space, cause disturbances in speech, a diagram of the speech region on the *surface* can give only a rough, approximate representation (Fig. 153). Only the names of the most important speech disorders can be marked upon it. If all the regions in which lesions produce difficulty in finding words (amnesic aphasia) were included, the region for speech would have to be drawn considerably larger.

The speech region, therefore, comprehends the lower, posterior part of the convexity of the left frontal lobe, the island of Reil, the front portion of the operculum of the anterior cerebral convolution (the Rolandic operculum), the posterior third of the temporal lobe, and one part of the lower occipital lobe.

These various parts of the speech region are not of equal importance.

4. *Complete Motor Aphasia*

(*Cortical Motor Aphasia of Wernicke*)

We must distinguish the *Frontal* speech region, the center nucleus[1] of which is the posterior two-thirds of the *inferior* frontal convolution (Pars triangularis and opercularis, the latter being *Broca's* region), but which, at least in many people extends to the lower third of the anterior central gyrus and the front part of the Island of Reil, probably even up to the *middle* frontal convolution. And, moreover, the anterior and posterior boundaries seem to differ somewhat in different individuals. Extensive lesions of this region cause disorder, which, clinico-psychologically, corresponds to the loss of motor word-components. The patients are still able to understand what is being said to them—a is retained—but can not find the innervation-complexes necessary to the utterance of words—that is, motor mnemic images are lacking. They are therefore unable to utter words (*word-dumb*, aphemic), or have left only a few fragments of speech. Because of the dependence of written speech on m, writing is lost or badly disturbed in the majority of people (see below)—except copying, which over o———gr (eye, hand) is possible without the use of (a—m). Even reading shows a gradual but very distinct deterioration. Reading aloud is impossible because of the inability to utter words, but the internal *comprehension of reading* is, for the most part, not seriously affected, although seldom, intact.

[1] Naturally, the word "nucleus" here, has nothing in common with the anatomical concept of "nucleus."

The resulting clinical picture is known as complete *motor* aphasia, consisting of:

Cessation of imitative speech.
Cessation of spontaneous speech.
Cessation of reading aloud.
Cessation of spontaneous writing.
Cessation of writing from dictation.
Difficulty in the comprehension of reading.
Comprehension of speech and copying are retained.

In *mental excitement*, the sufferer from motor aphasia (in contrast to the anarthric) at times correctly utters a word or even a sentence which as a rule he can not bring to utterance. The speech of many people thus affected shows improvement when singing. (Speech as an expression of the emotions may be partly retained, when as an expression of thought it is lost. Thus the motor-aphasic may swear roundly.)

For the retrogression of motor aphasia, and the peculiarities of returning speech, cf. page 529.

For clinical pictures approximating complete motor aphasia, cf. under insular aphasia, page 530.

5. *Complete Sensory Aphasia*

(*Cortical Sensory Aphasia of Wernicke*)

On the other hand, extensive focal injury of the *temporal* speech-region (the posterior third of the *superior temporal convolution* with the *temporal transverse convolution* and the adjoining anterior part of the supramarginal gyrus) produces *sensory* aphasia (cf. also pages 531, 534, 536).

In sensory aphasia, the memory of word-sounds (a) is severely impaired. Consequently, the main symptom is *word-deafness*. The patients hear, but fail to understand what is said to them. And since the sound of the word is not grasped, of course the meaning of the word is not understood. To grasp the sound of the word, more is needed than to hear each separate tone. Besides the formation of letters and syllables from the tones, and the ability to grasp them, the co-operation of the *memory of word-sounds* is particularly necessary.

Since, as we saw, acoustic components play a great part in speech, it is disordered by injury to them.

There is not indeed a cessation of speech, as in motor aphasia, but there are many derangements, and much confusion of words, syllables, and letters, known as *Paraphasia*.[1]

If the patient instead of knife says hammer, he has *verbal* paraphasia; if,

[1] In the derangements of the sensory-aphasic patient, "getting stuck" plays an important part.

instead of knife, he says life, he has *literal* paraphasia. If the latter is highly developed, the result is an incomprehensible jargon: a *jargon-paraphasia*, where one can understand, only by conjecture, what the patient means.

The degree of paraphasia varies greatly.

Many patients, suffering from sensory aphasia, can, by circumlocutions, avoid the words they do not command, and utter a lot of phrases, which while correct in themselves, are conspicuous by their lack of concreteness. Then the serious disturbances in speech appear only when one lets them name concrete objects (knife, chair, window, etc.).

The sensory aphasic in contradistinction to the motor aphasic, even in an advanced degree of speech-disturbance, commands a great number of letters and syllables, the expression of which causes him no trouble; but he does not always use the fitting ones, and often combines them so confusedly, as to be understood only partially, or not at all.

Such a patient is even usually *talkative* (scarcely realizing that his speech is unintelligible), while the sufferer from motor aphasia generally remains silent, hesitating to attempt speech (noticing his errors because of the intact acoustic word (a)).

Since written speech depends on a, it is disturbed even in sensory aphasia, the writing being, for the most part, better than in the case of motor aphasia. In sensory aphasia, the patient writes *paragraphically*, i. e., confusing the letters, as he does in speech, whereas reading is more affected than in motor aphasia. Alexia persists, more frequently only severe dyslexia. Reading aloud is paralectic, as speaking is paraphasic, and even the *internal* comprehension of speech is, for the most part, seriously disturbed.

The clinical picture of sensory aphasia, is therefore, as follows:—Cessation of speech understanding, or serious disturbance.

Spontaneous speech: paraphasic in varying degrees, with comparative wealth of syllables, often indeed of words.

Imitative speech: cessation or serious paraphasia.

Writing: paragraphic (spontaneous and dictated).

Copying: retained.

Reading aloud: paralexia.

Understanding of reading: seriously affected.

The main differences, then, between motor and sensory aphasia are:

	Motor Aphasia	Sensory Aphasia
Speech:	Lost.	Abundant, but paraphasic.
Understanding:	Retained.	Lost.
Understanding of reading:	Slightly disturbed.	Seriously disturbed.

Now, in sensory aphasia, *word-deafness commonly departs after some months* (by substitution of the right hemisphere). By strict tests, however, defects

in the understanding of words may be found years later. Paraphasia, paragraphia and paralexia are usually more lasting. Therefore one may not always expect *a high degree* of word-deafness from an old injury in the left temporal lobe.

While, from mere destruction of the *left* temporal speech region, not loss of speech, but paraphasia results, an additional focal injury in the *right* temporal lobe may cause *word-deafness*. This word-deafness happens in an entirely different manner from that of motor aphasia: all stimuli coming from the outside have been withdrawn from the motor-word center. In these two-sided focal injuries in the temporal lobes, the word-deafness remains stable, and should these focal injuries occur in certain places, complete cerebral deafness persists. Practically, therefore, temporal word-dumbness is differentiated from motor aphasia by its concomitant word-deafness, even common deafness. (Cf. also pages 534 and 356.)

The word *dumbness* of *motor* aphasia resulting from extensive frontal focal injury, is quite *lasting*. But even in this case, after considerable time—though in many individuals *never*—a certain retrogression may take place; after some months when the focal injuries are small, or when there are favorable compensating conditions.

First substantives and separate infinitives return, and adjectives without particles, without inflection and declension, one after the other. (Agrammatism or Telegram speech, i. e. Took walk, dizzy, sick, fall, hospital—as the narration of the course of his illness.)

By this telegraphic style, the speech of motor aphasia, after a certain retrogression, is distinguished from the speech of the sensory aphasic, which commands the inflexions and *forms* of speech, uses many well-built circumlocutory phrases, but can not find concrete nouns and verbs.

Furthermore, the former is distinguished by the *continuing difficulty in articulation*. In improvement in motor aphasia, imitative speech returns before spontaneous speech, and can become fairly good, though spontaneous speech long remains inadequate.

6. *Total Aphasia*

More frequently than a lesion confined to the *frontal* or *temporal* speech region, we find, as a result of the arterial distribution bringing all the blood, in the whole region of speech, through the *Art. foss. Sylvii*, lesions, that affect both regions, and therefore causing *total* or almost total (motor and sensory) aphasia. Following the retrogression of the word-deafness, one sees, years later, a clinical picture, in which the symptoms of *motor* aphasia predominate. Word-dumbness conceals the paraphasia; the disturbance of speech understanding is no longer very serious.

Writing and reading, since a and m are affected, are very poor. Hence

it happens, that old cases, in which the lesions occur in both speech regions, are often classed clinically only as motor aphasia. *In these cases, disturbances in speaking, writing and reading, are particularly stable.*

This clinical picture in its entirety of motor and sensory aphasia was named by Wernicke "*cortical.*" He assumed that the cortex was the seat of the mnemic images in question, and that only the *loss* of mnemic images gives rise to the complete symptoms portrayed. Naturally, this must not be interpreted so literally as to mean that the lesion must affect only the cortex. This seldom happens; usually, not only the cortex but also the underlying white matter is destroyed.

This, however, makes no change in the clinical picture, since the loss of the projection and association fibres that run through the white matter, can cause no additional disturbances, when once the center itself is destroyed.

7. *Insular Aphasias*

Between the frontal and temporal lobes lies the insular (Island of Reil) speech region. Focal injuries in the Island of Reil give rise to very different clinical pictures, which to this extent, resemble *motor aphasia,* in that they disturb the expressive phase of speech. But on the other hand, they injure the understanding of it only under circumstances to be more definitely described. They are:

1. Possibly by injuring the *cortex* of the insula, since, as we have mentioned, the front part of the insular cortex may belong to the motor speech center.

2. When they lie subcortically *between the cortex and the lenticular nucleus,* by cutting through essential bundles of white fibres, which connect a with m, namely, the external capsule and the extreme capsule. This interruption of part of the paths a—m causes difficulty in finding words, paraphasia, and injures imitative speech, but that less than spontaneous speech, since this simplest function of speech is naturally capable of most resistance. As long as some part of the connections a—m are retained—all of which by no means pass through the insula—imitative speech is possible, if not wholly intact. This is directly contrary to the older hypothesis, which held that in cases of insular injuries, it is precisely imitative speech that is the most seriously disturbed. Because of the dissociation of a and m, written speech suffers more or less, since, in the majority of people, it is dependent upon the internal word's being intact (a—m).

3. Frequently so-called focal insular injuries extend into the white matter of the third frontal convolution and the Rolandic operculum. In this case, we have besides the breaks (mentioned under 2) between a and m: 1. a break in the left centrifugal projection path to the bulbar cells; 2. interruptions in the fibres that pass from m to the Rolandic operculum; 3. of the

corpus callosum fibres; 4. destruction of the Fasc. arcuatus (cf. page 471), another very important connective path between a and m.

Hence, a break between a and m, would, on the one hand, be more vital than in two (2); on the other hand, a subcortical break of the pathways leading from m to the periphery exists. Therefore one finds clinical pictures closely approximating total motor aphasia: very poor, almost destroyed spontaneous speech, almost no imitative speech, and disturbances in written speech. Such insular focal injuries are of frequent occurrence.

4. Often the focal insular injury extends posteriorly down into the white matter of the temporal lobe, and then it may injure the understanding of speech.

From this it is clear, that focal insular injuries may have different effects, according to their situation and extent; they all cause, however, greater or less disturbance in the expressive phase of speech.

8. *Pure Word-dumbness*

Pure Motor Aphasia ("Subcortical" of Wernicke)

Besides the total motor aphasia (cortical of Wernicke), described above, another form occurs, but more seldom, *pure word-dumbness* (subcortical form of Wernicke). In this form only expressive speech, that is, spontaneous and imitative (by imitative is meant ability to repeat the words of others), is lost, whereas written speech (writing and reading) is retained. In some people, as has been mentioned before (page 524), written speech is more independent of motor-word memories, and can be maintained by the acoustic word alone. In this way, the rarer cases, in which the lesion is the same, as in total motor-aphasia, and yet written speech is retained, are to be explained. The majority of people, especially of the uneducated have to spell the word internally in writing (and reading), others (especially *those well versed in writing and reading*), need the motor word less frequently or not at all. The latter, in case of cortical focal injury in the anterior speech-region will show pure word-dumbness.

This would be pure word-dumbness by destruction of the motor speech center (*first* form of pure word-dumbness).

In the majority of people the clinical picture of pure word-dumbness is brought about by the fact that the motor center for speech is altogether or largely retained, but cut off from the operative muscular apparatus of speech. Under such conditions the motor images are spared, and remain at the service of the acts of reading and writing, but the escape of stimuli into the bulbar cells is prevented. In this case internal speech is retained, the memory not impaired, so that we dare scarcely call it aphasia. But since there is as yet no question of an injury in the outgoing neuro-muscular

speech apparatus, the disturbance belongs in no case to anarthria, but, as a break between the memory apparatus and the projection apparatus, lies between the two. Now, since the "purity" of this word-dumbness is always only approximate, and as a slight admixture of disturbances of internal speech can almost always be proved present, these cases are usually and justly classified under the aphasias.

In the belief, that in pure word-dumbness, the cortical part of the center for motor images is retained, and only the white matter and fibres running to the speech centers are interrupted, *Wernicke* called this form "subcortical aphasia." Since the older hypothesis of a direct path of projection (speech path) from the motor speech center to the bulbar cells has become doubtful, and it is far more probable, that from the motor speech center, the stimuli reach first the cortical center of the hypoglossal and facial nerves ("μ" in the scheme), *on both sides* in the inferior third of the frontal convolution (Rolandic operculum), and from there only to the bulbar cells, one should rather think of a break in these connections between the motor speech center and both sides of the anterior central convolution, which can actually be caused by subcortical focal injuries in the third frontal convolution and the adjacent white matter. (Break in the pathways from m to the left side of the Rolandic operculum and in the *corpus callosum paths* to the right side of the Rolandic operculum.)

The old conception of subcortical aphasia—better replaced by pure word-dumbness—may not find anatomically a strict application in words: the meaning is that a break in the paths between the motor speech centers and the muscular apparatus of speech is concerned. Naturally a subcortical focal injury outside of *these* connective paths, can interrupt so many association fibres of the motor speech center with the whole cortex, that the motor speech center is almost isolated, and therefore almost as functionless as if it were destroyed. Such a subcortical lesion would give a clinical picture of *total* motor aphasia. On the other hand, a subcortical lesion, according to situation and extent, may be of such a nature, as to spare partly the fibres passing from the speech center to the cortical centers of hypoglossal and facial nerves on both sides, perhaps the commissural fibres on the right passing through the corpus callosum, and then a subcortical focal injury would not cause even pure word-dumbness. Finally a focal injury may destroy part of the *cortex* without wholly incapacitating it, but with an extension into the white matter, break the connections with both sides of the Rolandic operculum. Then this focal injury, *partly cortical* and partly subcortical, would have a similar effect as a pure subcortical break of the connections named. It is evident, that the conceptions of cortical and subcortical aphasia are not anatomically to be taken literally, but mean as much as *incapacitation of the cortical speech center itself and a mere break between this and its subordinate centers*. In reality, many different combina-

tions of lesions may form one or the other clinical picture. But it has been proved, that in the case of extensive destruction of the cortex the usual result is the clinical picture of total motor aphasia, whereas pure word-dumbness results, in the majority of cases from subcortical lesions.

Excluding that minority, whose written speech is independent of the motor speech center, we may say: lesions, that spare wholly, or partly the cortex of the frontal speech regions, cause only the loss of spoken speech, and spare written speech. (Pure word-dumbness, second form of pure word-dumbness.) Those lesions, however, that to a great extent destroy the cortex of the frontal speech region, usually affect written as well as spoken speech. A lesion, moreover, that breaks the left cortical bulbar tract, in addition to a second lesion which breaks the commissural paths between m and the right Rolandic operculum, likewise causes pure word-dumbness.

9. *Pure Word-deafness*

Likewise we have, beside total *sensory* aphasia, an isolated or pure word-deafness (*Lichtheim's* Disease; subcortical sensory aphasia).

In this disease, only comprehension, and as a natural consequence, imitative speech and dictated writing are lost, whereas speaking, writing, and reading are retained. Paraphasia, paragraphia, and paralexia are wanting to make this total sensory aphasia. That a focal injury, which destroys the sensory speech center, causes pure word-deafness (because an *individual* independence of spoken and written speech of auditory images is in question, corresponding to the oft discussed relation when motor-word images are disturbed) is necessarily a thing of rare occurrence.

Even the dependence of *written* speech on the acoustic word has fewer exceptions than that on the motor-word. In a case of pure word-deafness, one must think of the persistence of the substratum of auditory word images, and of mere exclusion of these from acoustic stimuli. This, as *Lichtheim* has taught, is caused in the first instance, by a subcortical focal injury in the left temporal lobe, situated so as to break the incoming of all acoustic stimuli to the left temporal lobe, and possibly, too, the corpus callosum connection between the hearing center of the right and that of the left hemisphere. Then the possession of auditory word images is established by the fact that reading, speaking, and writing are intact, whereas the exclusion of these auditory word images from all acoustic stimuli, causes the loss of speech-comprehension. A second possibility as to the establishment of pure word-deafness occurs, if with *Flechsig*, one separates the acoustic *projection* center sharply from the *mnemic* acoustic center (cf. Fig. 131). Then, the destruction of this center (i. e., particularly of the transverse convolution in the temporal lobe) deprives the left hemisphere of acoustic stimuli and hence cuts off auditory word images from the periphery, whereas they could

still develop their *intercortical* effectiveness in speaking, writing, and reading. This would be pure word-deafness due to *cortical* focal injury (in the transverse convolution). It is doubtful whether pure word-deafness could arise from a break in a path alpha—a, since it is doubtful, whether alpha and a are connected by a *complete set* of association fibres.

Pure word-deafness persists, provided the right temporal lobe does not, in the course of time, also take the place of the left, which, in the case of subcortical focal injuries, often fails to happen. Here, too, as in total sensory aphasia, the comprehension of the spoken word is already lost. Tones are heard, but only as a strange confused noise. If the comprehension of the *spoken* word is lost, naturally the *meaning* of the word is not grasped, i. e., the comprehension of the meaning of the word is wholly lost.

To establish a case of pure word-deafness, it must be proved that hearing is possible. If hearing is seriously impaired on both sides by disease of the labyrinths or of the auditory tracts or centers, the comprehension of speech may be lost as *a result of defective hearing*, alone. Then we have pseudo-speech-deafness, a mere consequence of defective hearing. Since *Bezold* proved, that total loss of the tone range b′ to g″, or serious weakening within this range, eo ipso, destroys comprehension of speech, one must prove not only the continuous range of tones but also that the patient hears with sufficient clearness within the range of the tones concerned. It is not sufficient to prove the existence of hearing, by received perception of noises, whistling, clapping of hands, ringing, etc., since if the tone-range mentioned above is lost, speech will not be understood, even if the hearing of the other tones of the entire range of tones is intact. Furthermore, to distinguish subcortical sensory aphasia from loss of comprehension of speech, due to general deafness, this fact serves—that the former is never *absolutely* pure, but that separate paraphasic or paragraphic symptoms prove that it is not a question of mere defect in hearing.

There is besides a still rarer form of speech-deafness, in which letters, syllables, and short words can be comprehended and repeated, but larger words and sentences are not understood, not even in the sound of the words. With this there is only very slight paraphasia, paragraphia and paralexia, so that this form (because of the small proportion of disturbances in internal speech) approximates pure word-deafness. A cortical focal injury in the speech center, which injures auditory word images less, than their communication with the acoustic stimuli, might be the cause.

10. *Transcortical Aphasias*

(*Including Amnesic and Visual Aphasia*)

By transcortical aphasias, *Wernicke*, indorsing the theory and classification of Lichtheim, understands those, in which the motor and sensory speech

centers themselves, and their connection with each other and with the periphery are retained, but one of the two is cut off from the perception area, i. e., from all the rest of the cortex. Psychologically, the word (a—m) is intact, but one of its components is cut off from B. The complex ear (a—m) mouth (Fig. 154) is therefore free.

W. thought this was caused by a break in the association fibres which converge from the entire cortex to one of both speech centers.

In his scheme, disturbance in a and m means cortical aphasia, a break in the pathway between m and a, subcortical aphasia, and a break in the paths from B to m or to a, transcortical aphasia (correspondingly motor or sensory).

Clinically this is marked especially, by the ability to repeat words (the comprehension of *spoken* speech is retained), inasmuch as the section ear—a—m—mouth is free, but that either the word can not be found from the meaning (transcortical motor aphasia, 6) or that the meaning of the word heard cannot be found (loss of comprehension of word *meanings*) transcortical sensory aphasia (3). The disturbances in writing and reading ascribed to the transcortical forms (spoken speech, dictated writing retained) are realized only approximately. The *clinical* reality of these transcortical forms, with this limitation, is certain: we often have patients, who can repeat words (sound comprehension retained), but who do not understand them readily (meaning comprehension lost) or have difficulty in finding words from their meaning. Often *these two* are associated: difficulty in the comprehension of the meaning of words and very great difficulty in finding words, though imitative speech is retained.

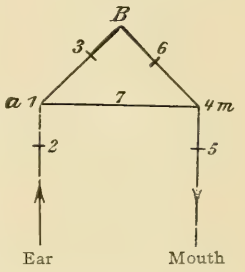


FIG. 154.—Wernicke's scheme. 1, Cortical sensory aphasia. 2, Subcortical sensory aphasia. 3, Transcortical sensory aphasia. 4, Cortical motor aphasia. 5, Subcortical motor aphasia. 6, Transcortical motor aphasia. 7, Conductive aphasia (clinically not confirmed).

But **anatomically,** the relations assumed by *Wernicke* are not to be realized in the simple manner, taken from the brain, as he supposed. It is not merely the case that, when imitative speech is lost, the focal injury is in the center, and when it is retained, that the focal injury lies in the tracts converging to the center. The principle, in Wernicke's supposition, that when imitative speech is lost, the real conveyors of the motor or sensory word are destroyed, and that when it is retained, they are retained, but can not be reached from the superior cortical seat of perception, holds good in part, but under very different anatomical conditions, and sometimes the focal injuries are so situated that one does not, without further difficulty, see this effect. The empirical conditions, and the knowledge that the direct path B—m is of no great importance, that moreover, spontaneous speech mainly uses the same pathway as imitative speech, namely the path a—m, necessitate a modification of this part of *Wernicke's* view. Retention and

loss of imitative speech depend on other conditions beyond the situation of the focal injuries, that is, on the *degree* of lesion. Imitative speech, naturally, is, on the whole, that *function of speech, most capable of resistance,* so that slighter lesions of the same region, the destruction of which causes total aphasia, permit of some imitative speech. This is true also of the *paths* that lead to m (a–m). In partial lesions, the powerful stimuli, which come from the periphery, are allowed to pass, while the weaker spontaneous stimuli coming from B are not.

The word *transcortical* includes a clinico-symptomatological, an anatomical and a psychological set of conditions, all three of which are not always fulfilled together. We shall use the word here *only in its clinico-symptomatological sense,* i. e., consider as transcortical those forms only in which *imitative speech is retained.*

This is not the place to discuss the value of the other meanings of the word transcortical.

(A) *Transcortical motor aphasia* is *here,* the loss or serious disturbance of spontaneous speech, associated with retention of imitative speech.

This is found in:

I. Slight injury of m itself, which destroys the possibility of stimulating the center of perception from the periphery (via a), without injuring those fibres in the center that convey the motor-word to such an extent that they can not be innervated by the *stronger* stimulus of the spoken word.

II. Slight injury of the paths from a to m, a case already mentioned under focal insular injuries (page 530) and of the accessory paths from B to m from the same cause.

III. When one has to consider the possibility, that the return of imitative speech has become a function of the right hemisphere.

In cases 1 to 3, imitative speech is *better* than spontaneous speech, but nevertheless very much impaired and faulty.

IV. The requirement of the retention of imitative speech is more strictly complied with in so-called *amnesic aphasia or verbal amnesia,* in which, it is true, even *spontaneous* speech is not wholly lost. It is clinically, and principally, a slighter form of transcortical motor aphasia, though the mode of its origin be other than the one accepted by Wernicke for the latter case.

There is great difficulty in finding words, but if the word is suggested, it is immediately recognized as the right one, and repeated with ease and correctness. In this verbal amnesia, it is most difficult to find nouns and verbs for concrete objects or actions, whereas in contrast to forms 1 and 2, abstract nouns and inflected parts of speech, particles, prepositions, etc., as well as inflections and declensions are retained. This form is found under *manifold conditions,* always when one of the stations by which the process of speech must pass, before reaching the motor center, is slightly injured. Therefore:

1. When the *sensory* (temporal) speech center is but very slightly injured (say, by moderate atrophy). This small injury does not destroy imitative speech and comprehension, but makes difficult the wakening of auditory word images from the perception of them (*the function most easily disturbed*).

2. When the connections between the sensory speech center (a) and B are slightly injured, because of which it is difficult to find words, while the meaning of the word heard is still comprehended.

3. When the perceptive region itself, that is, the entire cortex is injured, even before the perception is so seriously disturbed, that verbal amnesia is lost in imbecility, the pathways to the sensory speech center are, in their very beginnings, injured.

In diffuse processes (paralysis, senile atrophy, arterio-sclerosis) frequently several of the conditions named under 1 to 3 are fulfilled, so that in those cases, amnesic aphasia is quite usual.

But, moreover, focal injuries back of a, which are not sufficiently *extended*, to produce a case of transcortical sensory aphasia (to be discussed later), reduce spontaneous speech to the lowest degree, without in the least affecting imitative speech.

The so-called *visual* (*optic*) *aphasia* is only a peculiar form of amnesic aphasia. According to its definition, objects seen can not be named, and those objects, that we know exceedingly well and mostly by sight, can not be designated in free speech. On the other hand, objects perceived by any other sense, for instance, that of touch, it should be possible to name. In reality, the most frequent form is an *optic-tactile* aphasia, i. e., neither by sight, nor by touch, is the name of an object found, though it may be easily found by ear. A trumpet, for instance, that has been seen and touched, can not be named, whereas as soon as it sounds, the name is immediately suggested. In this visual aphasia, it is generally an extension of amnesic aphasia, that we find, in which, the inability to name objects (in contrast to common parts of speech and abstract nouns) is more than usually conspicuous, in which the perception of the object creates less favorable conditions for naming it, than exist in free speech, and naming by acoustic characteristics proves to be easier.

This hall-mark of amnesic aphasia is found especially in cases of *abscesses in the temporal lobes*, which mostly lie at the base of the temporal-occipital lobes, furthermore in other injuries in the *connecting pathways between the temporal and occipital lobes*, even *in the occipital* lobes themselves. They have this in common, namely, that they interrupt connective paths between a and the occipital lobe, the seat of the essential optic components of perception.

Amnesic and especially optic aphasia is *transcortical*, even in *that* meaning of the word that has been heretofore neglected: the hindrance in speech does not lie in the word centers themselves (a or m), nor in their connections with

one another and the periphery, but beyond the centers, on the path that must be transversed by the stimulus before it can reach the word centers from preception.

Since a is for us an intermediate station for the stimulus from C to m, in this sense every hindrance in expressive speech caused by a lesion in a is transcortical, especially for instance, the word-dumbness due to bilateral injuries in the temporal lobes (see page 529). In this case, however, imitative speech also is lost so that the distinctive feature that *clinically* and particularly characterizes transcortical aphasias, *is absent.* This difficulty we may only point out here.

(B) *Transcortical Sensory Aphasia.*—We have seen that an injury to the connective pathway between a and B, that is, a *partial* interruption only, causes transcortical motor aphasia, seriously hindering spontaneous speech (that is the stimulus in the direction B to a), but does not destroy the pathway in the opposite direction a to B: the connection of the percept with the word, the comprehension of the meaning of the word.

If, however, a *total* interruption occurs in the paths from B to a, as opposed to partial interruption, or if B itself, is seriously injured (asymbolia in the older sense of the word) we have *transcortical sensory aphasia:* imitative speech is retained, often occurring *irrepressibly* (*echolalia*), but comprehension of what is spoken is lost. This happens, because no concept joins the auditory word image evoked in a, but the stimulus, to which the path to B is closed, flows out into the free path a to m of imitative speech. Anatomically transcortical sensory aphasia is shown frequently by marked atrophy in the *neighborhood of the first temporal convolution*, that is, in the second and third temporal convolution and even further back (marked atrophy in the white matter of the temporal lobe). It occurs often side by side with general atrophy and corresponding injury in the perception centers. (Asymbolic disturbances in the older sense.) Several focal injuries, moreover, in the posterior half of the brain can bring about this isolation of a, which spares only the connection with m.

Now, it must be remarked, that, if only a partial interruption in a—B, brought about almost complete loss of expressive speech, a greater interruption would certainly alter expressive speech in a considerable degree. As a matter of fact, we have almost always associated with transcortical sensory aphasia a serious disturbance in spontaneous speech. Hence transcortical sensory aphasia, as a rule, is associated with transcortical motor aphasia; only the *sound* of the word is comprehended, and parrot-like imitated, but neither is the *meaning* understood, nor spontaneous speech to any extent possible. A complete restriction to the symptom-complex actually found in transcortical sensory aphasia (i. e., imitative speech, but lack of comprehension in the meager spontaneous speech) is possible only in a person in whom, exceptionally, the pathway B—m is particularly efficient.

Summary of Temporal Lesions.—To summarize what has been said about sensory aphasic disturbances, an interruption of the path leading to the left auditory center (including the corpus callosum connection), as well as one in the left auditory center itself (middle third of the superior temporal convolution and transverse convolution), creates *pure* word-deafness. A destruction of the "sensory speech center" (posterior third of the superior temporal convolution and that part of the supra marginal gyrus, lying directly behind it) causes word deafness combined with paraphasia, paragraphia and paralexia. Where there is deafness to the word-*sound*, we find, in consequence deafness to the word-*meaning*.

A total isolation—with the exception of the connection with m—of the sensory speech (serious atrophy of the whole temporal lobe, extensive focal injuries in the depths back of and around the sensory speech center), destroys only the comprehension of the meaning and not that of the sound of the word, which depends on imitative speech, and at the same time produces the greatest difficulty in finding words. But if the sensory speech center or its connection with B is but slightly or partially disturbed, only amnesic aphasia results (in the second case, especially, optic aphasia), whereas comprehension of word sound and meaning are retained.

It is now self-evident, and this is true of all cerebral lesions, that diseases affecting the left temporal lobe, whether they be neoplasms or focal injuries of vascular origin, only exceptionally, restrict their destructive and far reaching consequences to the substratum of one element or an element connective, as sharply differentiated in the scheme. According to their various situations and extent, and to the participation of many pathways, represented by a *single* line in the scheme, various combinations of symptoms appear. Loss of comprehension of word-sound and word-meaning, difficulty in finding words and paraphasia, disturbances in writing and reading, combined in varying degrees, form the typical clinical pictures of the diseases which can be classified only approximately according to theoretically developed forms. Pure word-deafness, total sensory aphasia, and "transcortical" sensory aphasia, are only three particularly striking types of sensory aphasia, the diagnosis of which has great instructive value.

In order to comprehend all real differences, one would, in each case have to take into account, the extent to which the cortex, the projection—association—and commissural fibres are affected, and would moreover, have to know clearly the function of each of them, a knowledge which we have unfortunately by no means attained.

Disturbances in Written Speech

(*a*) *Alexia and Agraphia;* (*b*) *Pure Alexia;* (*c*) *Pure Agraphia*

We noticed that written speech was usually affected with spoken speech: writing, seriously in both main forms of aphasia; reading, seriously in sensory,

less seriously in motor aphasia. Only in the so-called pure forms was written speech retained intact.

There are disturbances in written speech, that are almost isolated, i. e., reading and writing are seriously impaired, while little harm is done to the other functions of speech.

Focal injuries, situated directly behind the sensory speech center, that is, in the angular gyrus and the *upper layers* of its white matter, cause *agraphia and alexia*, associated with but slight symptoms of paraphasia, and difficulty in finding words. Those focal injuries, however, which from the medial surface of the center extend forwards into the *deep* white matter of the angular gyrus, cause, especially, *pure alexia*, i. e. writing is retained, reading lost. With this right hemianopia is almost always associated, whereas superficial focal injuries in the angular gyrus, which cause the loss of both reading and writing, often do not produce hemianopia.

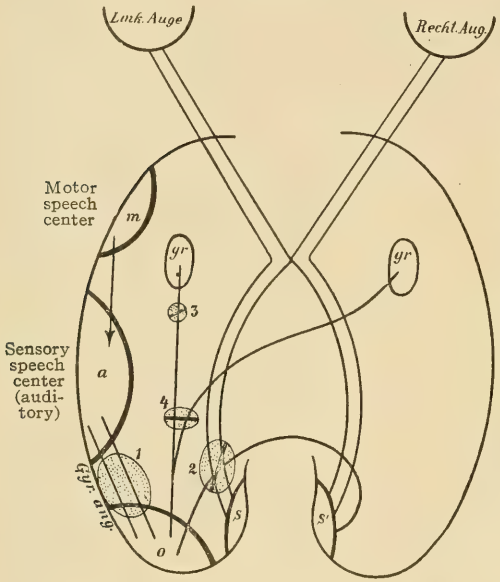


FIG. 155.—(Explanation in the text.) Focus 1 causes alexia and agraphia. Focus 2 pure alexia with hemianopia. Focus 3 pure agraphia of the right hand only. Focus 4 pure bilateral agraphia.

The simplest explanation of these statements, is as follows:

The optic radiation runs through the deep white matter of the angular gyrus, outside of the posterior horn through the white matter of the occipital lobe, to the calcarine region, the left and right visual center (s and s'). From here pass connecting fibres (not shown in Fig. 155) to the convexity of the occipital lobe, the seat of visual images (form memories) (s—o). These are registered *mostly* in the left, but also in the right occipital lobe. The connective fibres from the right occipital lobe pass through the splenium to the convexity of the left occipital lobe (s'—o). From the sensory speech center an association path passes to the convexity of the occipital lobe (a—o). From o to the hand center (gr) passes the path by which visual directions are sent to the hand (Fig. 155).

Now, if a focal injury destroys (1) the angular gyrus, starting from the convexity, it interrupts the path a—o. The consequence is, that the sound pictures of the letters aroused in a, do not awaken the corresponding visual images in o, which is necessary for writing; whence agraphia. In reading, the opposite takes place; the stimuli of both fields of light are not carried

over to a, so that images of letters fail to call up their sounds (for the sake of simplicity, we pay no attention to the secondary connective path o—m), and what is seen, is not understood; whence alexia. Because of the interruption in an important path from o to a, there results difficulty in finding words, and because of the proximity of the focal injury to a, there is frequently slight paraphasia. But if the focal injury lies in 2, it interrupts, on the one side, the optic radiation, hence right hemianopia, on the other, the commissural paths from the right occipital lobe (s′—o) so that no optic stimulus reaches the left o. The visual stimuli, that reach only the right occipital lobe, do not reach a, and therefore call up no auditory images, so reading is lost. But focal injury in 2 does not break the connections of a and m with o, nor those between o and gr, so that there is no reason for the loss of writing, hence pure alexia.

If a focal injury (3 and 4) interrupts the path from the left occipital lobe to the center for the right (or the left) hand, pure agraphia appears, usually in both hands, but under certain conditions, only in the right. Pure agraphia is also often a partial symptom of apraxia of the hand affected.

If one considers, that in the brain, it is not, as in the scheme, a matter of linear connection, but of numerous and various connections of fibres, that these pass to the other categories of fibres, on various levels and in various situations, that the size, extent, direction, form, and number of the focal injuries create the most varied combinations of interruptions, it is then clear, that the above linear scheme is merely a simple chart, but that, in reality, manifold combinations and varieties may appear. Suppose for instance, that focal injury 2 extends an outshoot so far laterally, that o—gr, too, is interrupted, then we shall have alexia associated with agraphia.

Pure alexia is caused by a focal injury in the region supplied by the posterior cerebral artery.

(b) Agnostic Disturbances

Word-deafness and reading blindness are really special manifestations of disturbances of *cognition*. The patient in reading blindness sees, but fails to understand what is seen. Perception is retained, but comprehension lost. Reading blindness is agnosia for written letters, word-deafness agnosia for word-sounds. In these receptive aphasic disturbances, it is a question of agnosia for conventional signs, for the symbols of speech. Speaking of agnosia, in the stricter sense, one means corresponding disturbances for the *things of the outer world*, namely, for all sense impressions, that are *not* symbols. (*Object agnosia.*) If say, a comb is seen, but its significance not recognized, because the complex of visual perceptions does not awaken those associations that normally appear, for instance, that the comb is the thing with which we comb our hair, that it belongs with the brush, etc., an agnostic disturbance is present. It is not a matter of making

the connection of the *symbol* and its import, but of the object's *real* bearing, origin, purpose, etc. According to the sense affected, one differentiates between acoustic, visual, tactile, gustatory and olfactory agnosia.

1. *Acoustic* agnosia or *mind deafness* is characterized by the lack of comprehension of speech sounds and all possible auditory stimuli, such as cries of animals, musical instruments, cracking of the whip, clinking of money, etc. It occurs associated with sensory aphasia, when the focal injury is in the left temporal lobe.

2. *Visual* agnosia or *mind blindness.* The patient can discuss the form and even the color of objects, but they are strange to him; he not only can not *name* them, when shown, as in visual aphasia, but can really not *recognize* them.

Mental blindness results from lesions in the occipital lobes and generally occurs when the focal injuries are bilateral. Usually by a lesion in the visual center, or optic radiation, half or at least a quarter of the field of vision is excluded, i. e., for this part of the visual field the person is, *perceptively* speaking, blind. The comprehension of the impressions received by the unaffected parts of the field of vision, is lost because of an interruption in the association fibres, which run from the visual center to the seat of visual form memories in the convexity of the occipital lobe, or because of the destruction of the center itself or its connections with the other cortical areas. This can be caused by various combinations of injuries, which affect the white matter and the convexity of the occipital lobe, and those parts of the parietal lobe lying directly in front of it. One of both injuries must always be *on the left;* focal injuries to the right occipital lobe do not, except in left-handed people, cause mental blindness. This proves, that the stability and delicacy of visual images in the left hemisphere are greater than those in the right. Indeed, in the majority of people the left hemisphere is so predominantly the seat of visual images, that, in them, an injury that is *only* left-sided effects a considerable degree of mental blindness. Mental blindness caused only by a left-sided injury, occurs, in such cases, in the following manner: destruction of the left visual center or of the optic radiation entirely cuts off optic stimuli from the right hemisphere (right hemianopia). The focal injury further breaks the corpus callosum connection between the right occipital lobe and the left hemisphere (splenium or forceps), whereby the visual stimuli reaching the right field of vision, cause perceptions, but are not understood, since they do not reach the seat of memories stored up predominantly on the left side. At the same time there naturally occurs pure alexia, since the mechanism of mental blindness, due to one-sided focal injury, operates as in pure alexia. Still other combinations of lesions are possible.

Usually perceptive and gnostic disturbances are found together, so that for one part of the field of vision there is blindness, and for that part of the field that has been retained, mental blindness.

As a rule, the acuity of vision is also impaired, often, too, there is some color blindness. Before one diagnoses mental blindness, it must, of course, be proved that these perceptive disturbances are not so weighty, that it is not due to the presentation of data, insufficient for the recognition of the objects. (Pseudo-mental blindness.)

In many cases of mental blindness visual memories are *lost*, in others, they merely can not be *aroused* by centripetal stimuli but the patient can somehow, by description, or drawing, indicate the possession of memory.

Or, although the visual memories are really aroused, the associated memories of the other senses, tactile, acoustic, can not connect with them. Often the reason of failure to recognize is that the *composition of the concept* of the object *from the various sensory data* is impossible. The conceptions, then, remain a chaos of simple impressions, which do not flow together to form concepts of concrete objects.

Usually, not only form memories, but also color memories are injured; the patient can not imagine the color of the frog, of the carriage, of the blood. Occasionally color memories are retained; but it is an exception when form memories are retained and only color memories lost. Usually, in mental blindness, memories of the spatial order of things suffer. The patient can not see "in his mind's eye," roads that were formerly familiar, and therefore easily loses his way.

Not to be confused with disturbance in color *memories* (loss of color images) or with disturbances in the *perception* of colors (color blindnes—tested by sorting colors), there is a disturbance in *naming* colors, occurring now with, now without the other two (inappositely named amnesic color blindness). In this case the patient can not give the right name to a color, just as he can not correctly name other things. (One of the phenomena in amnesic and so-called visual aphasia, that shows itself especially in names of colors and persons.)

Amnesic aphasia is usually associated with mental blindness and alexia, eventually also with agraphia.

3. *Tactile Agnosia* ("*Touch Crippling*").—In tactile agnosia the patient does not recognize the object touched in spite of the presence of various and sufficient incoming sensations (perceptions of touch, location, movement, etc.). For example, forms are not recognized (astereognosis). Nor do perceptions of moisture, cold, velvety softness, etc., lead to correct conclusions.

There are three kinds of tactile agnosia:

1. Either separate impressions are not correctly correlated, though tactile memories are retained, that is, they do not correctly evoke the latter, or

2. Tactile memories are lost, or

3. The association between them and the visual, auditory, etc., memories is broken.

Anatomically, in tactile agnosia, focal injuries exist in the *middle third of the posterior central convolution*, or behind it, in the parietal lobe, the latter being especially concerned in the third associative form.

Not to be confused with tactile agnosia, there is that failure to recognize through touch, which is caused by *severe disturbances in sensation* and by lesions of sensory paths. In this case we find not an *agnostic*, but a *perceptive* disturbance, and one should distinguish also, similarly caused disturbances in form recognitionas, as perceptive astereognosis, from tactile agnosia (see page 506).

4. Agnosias in the spheres of *smell and taste* are distinguished with difficulty from the corresponding perceptive disturbances and have been little studied.

The agnostic (dissolutory) disturbances that have been heretofore discussed, rest on the fact, that the members of the series of ideas essential to cognition, split into their *separate sensory components*, or are injured by the destruction of such a component; for instance, the optic. Frequently, however, a disturbance in recognition is caused by the fact, that ideas, *intact* in their *sensory* elements, are combined in the wrong order, that perhaps the connection between the partial conceptions and the whole concept, or the correct association of cause, purpose, or characteristics with an object is lost, that is, those associations, necessary to recognition, are injured that do not precisely concern the connection of the *sensory* elements (*ideational* agnosia, counterpart, and frequently companion of ideational apraxia).

Various associative and attentional disturbances thus affect the train of ideas. The object is, for example, not recognized, because a "persevering" idea, or an accidental sensory impression, or an emerging immaterial side issue interposes and turns aside the train of ideas.

These ideational agnosias occur especially with diffuse mental processes, furthermore as general effects of focal injuries, hence often accompany the focal symptoms, possibly occur even as direct focal symptoms. They present those disturbances in cognition, that one generally characterizes as "psychic."

If optic, tactile and acoustic agnosia are associated, we have *total* agnosia, used in the older sense of total *asymbolia*. This presupposes such extensive injuries, large lesions in both temporal, parietal, and occipital lobes, that *memory pictures* themselves and their *connection with understanding* are virtually destroyed. Then there is entire *loss of comprehension*.

(c) Apraxic Disturbances

1. *General Remarks*

In brain diseases, there is often inability to move the limbs, so as to carry out the movements intended by the patient, so that even very familiar and

thoroughly learned movements can not be made, though neither palsy nor ataxia is a sufficient cause.

For the patient can *occasionally* contract all muscles; he raises, lowers, bends, stretches arm and hand often vigorously and once in a while executes complicated movements, and is thus differentiated from the paralytic; but just when he wishes to make a certain movement he is unable to do it. Other apraxics can carry out such simple purposive movements, but the still relatively simple movement complexes, such as greeting, beckoning, threatening, lighting cigars, sealing letters, pouring out water, etc., they can not execute. Objects are wrongly used, or the patient stares at them, being at a loss as to what to do with them.

The limbs then can not be made to serve the purposes of life. As in aphasia, the unparalyzed muscles of tongue, lips, and palate, can not be so directed as to produce the word in mind, the apraxic can not make his paralyzed hand execute certain movements. The expressive-aphasic disturbances present only part of the symptoms of apraxia, just as the receptive-aphasic present part of those of agnosia, and it is only from historical considerations that aphasic disturbances are treated separately.

The inability to perform purposive movements, and purposive groups of movements in the every-day affairs of life, arises from very different causes.

Partial recollections (i. e., kinetic or optic) for combined movements may be lost, or difficult to rouse, or the connection among the partial components may be lost, owing to an interruption in the fibres, or the regular outlet of the mental process that *prepares* complicated movements, step by step, has been affected by various disturbances (consisting not exactly in loss or splitting of partial memories) of mnemic, associative, attentional kinds.

The apraxic suffers *not* from a disturbance of those *lower* co-ordinations of muscles, known as *ataxia*, which is caused by severe sensory disturbances.

Taxia so regulates the working together of muscles, that the limbs, without wavering, on the shortest path, traverse the road to the goal. But *which* road is to be traversed, by *which members or parts of members*, *in what co-ordination or sequence*, *which object* is to be the starting point, in order that the desired form of motions, and that demanded by the purposes of life, may result, and how, in the use of taxia, all necessary simultaneous and successive innervations shall occur—in these questions, taxia says nothing, but praxia rules.

For practical differentiation of the ataxic, there serves among other tests, this—that the ataxic performs *inexact* movements, while the apraxic (for exceptions see below) performs frequently movements—quite different from those desired, that, on the other hand, while the movements of the apraxic do not correspond to the intention, they appear perfectly co-ordinated, viewed in the light of *another* purpose. He may write the wrong letter, that is, in itself correct, or he places the comb behind his ear with the muscular move-

ments perfectly suited to *this* act. Those apraxics, who do not know how to handle objects, were formerly usually classified with the *agnostics*. The word apraxia was formerly employed when some one used an object incorrectly. But it was thought that he did so from *mistaking* the object or failing to recognize its *use*. As *one-sided* apraxics prove, a man can, however, recognize the object, know its use, and yet be unable to use it.

In order to diagnose apraxia, it must be proved that the patient *recognizes* the object. *At first sight*, he seems to be *mistaken*, as when he makes motions of smoking with a tooth-brush, as if it were a cigar.

The pre-essential condition of purposive movement, as, say, the lighting of a cigar, is that the details of this movement arise internally, in correct sequence, and in relation to the *right object*, so that the train of ideas corresponds to the sequence of the necessary partial movements, and their relations to objects (in very frequently performed acts, the *subconscious* cerebral equivalent of this train of ideas). This is the *ideational* "plan" of movement, which determines which paths, in what sequence, are to be run over in relation to what object. In order that the corresponding movement may be actually reached, directions corresponding to the ideational plan, must go to the motor center of the *executing* limb, i. e., kinetic-innervational memories must be awakened which release the real innervation.

If the ideational "plan" for movement is wrong, we speak of *ideational* apraxia; if the disturbance is only in the transmission of ideational plans to the special kinematic of the executing limb, we speak of motor apraxia (in the broader sense).

We begin with the latter.

2. *Motor Apraxia*

Two subordinate forms are to be distinguished:

1. Motor apraxia in the *narrower* sense or ideo-kinetic apraxia. Here the limb-center with its kinetic-innervational memories is itself retained, but through breaks of countless connections with the other brain centers the transmission of the "plan" of the movement to the limb-center is hindered. The disease tears asunder ideation (plan of movement) and limb kinematics. It must be assumed that in the sensory-motor limb center, not only the seat of innervation, and the substratum of synergies is laid, but also the substratum for certain much practised simple movements, the building stones of complex purposive movements, as blowing, whistling, winking, writing a letter of the alphabet, etc.

These simplest actions do not need directions from the brain as to every step of their movement, but are a special possession of the limb centers, as are the synergies. The retention of this special possession of the limb center is shown clinically by the fact, that occasionally well-formed movements are executed, i. e., a correct letter written, hands clasped, etc., but not *at the*

proper point, not when the patient wishes and is determined to do it, because the necessary co-operation of the limb-center with the brain is lacking.

2. If on the other hand, from a lesion in the limb-center itself *that does not go so far as to paralyze it*, the special possession of the limb-center of kinetic memories, be injured, the seat of innervation can still obey, in part, the orders of the brain; but the loss of this possession, given to the limb by long practice, causes all movements to become rough, inexact, uneconomic, like those of a man who is attempting them for the first time, with the result that such movements that are not planned in the rest of the brain, but exist only as kinetic memories of the limb-center, as blowing and whistling, can not be performed at all. This subordinate form of motor apraxia may be called limb-kinetic apraxia.[1] In so far, however, as co-operation between limb-center and the whole brain exists, usually through the mediation of this specialized possession, its injury will cause "derailments," which resemble those caused by breaks in the paths running to the limb-center, namely, the ideo-kinetic.

(α) *Clinical Aspect of Ideo-kinetic Apraxia (Motor Apraxia Par Excellence)*

It is confined to separate limbs, often half of the body. (For exceptions see below.)

A train of simple movements is, *on occasion*, quite correctly performed: the patient closes his hand correctly when he grasps an object, but is unable to do the same thing when he wishes to make a fist; in writing he writes the wrong letters, though each is correct in itself. Beyond this *occasional* occurrence of correctly executed movements, however, even the very *simplest movements fail*, when the stimulus comes from this part of the brain from which the limb-center is cut off. The patient can not copy a vertical or horizontal line that has been drawn for him, nor open his fist when he is told; more complicated movements, pouring out water, striking matches, etc., are out of question. The following kinds of false reactions occur:

1. Movements that resemble no purposive movements, such as beating the hand, spreading the fingers (so-called *amorphous* movements).

2. *Parapraxic movements*—winking in place of threatening, touching the ear instead of the nose, etc.

3. The movement is transferred to an *entirely* different musculature—standing straight and rigid, in place of extending the hand. This simulates real loss of movement.

4. Often motor indecision and *real loss of movement* take place. In many miss-reactions, there is shown a strong tendency to *fixation* of ideas (perseveration), namely, the intended movement is not executed, but the preceding one repeated. By combination of such "persevering" movements with components of the new purposive movements, very curious bastard

[1] This conforms, at least so far as definition is concerned, with *Meynert's* motor asymboia.

formations arise. The fixation of movements is probably not the cause, but the result of the right movement not being executed. Ideo-kinetic apraxia is shown even in *imitative* movements.

5. Secondary disturbances of the nature of ideational apraxia, to be described below, appear.

(β) *Clinical Aspect of Limb-kinetic Apraxia*

Since very extensive focal lesions in the limb-center lead to *paralysis* (which conceals the loss of limb-kinetic memories), it occurs mainly in discrete processes (arterio-sclerosis, senile atrophy, progressive paralysis). The coarse movements are awkwardly, inexactly performed and resemble those in cerebral ataxia. They are distinguished from them in this, that for many *more delicate* actions, sewing, blowing, whistling, not even a tendency towards the action is to be distinguished. The movement shown in these actions, is not only an ataxic distortion of the correct one, but is wholly lost, or bears not a trace of resemblance to the original form. Here, too, secondary ideational disturbances may occur.

3. *Ideational Apraxia*

Here manifold disturbances even in the ideational plan, are concerned—due to flaws in memory, attention and association. The patient omits parts of an action, performs them in the wrong order, or makes the right movement towards the wrong object. Such a strange confusion of movements results, that the patient, e. g., instead of striking the match, places it in his mouth, next to a cigar, or attempts to cut off the end of the cigar, by inserting it between the two parts of the match box. In sealing, he puts the seal in the fire, and then impresses it on the wax. The errors look like results of absent-mindedness; generally the association bond between correct and incorrect movement can be proved. Single movements are quite correct.

Ideational apraxia appears only in complicated movements, or, at least, grows with the degree of complication. *Imitation* of short movements is retained, since here the plan of movement is given the patient from without. Only that the form of movement does not spontaneously occur to him. (If the latter disturbance predominates, and other ideational derangements appear in a small degree, or not at all, as is often the case, one may call it *amnesic apraxia*.) It is, in general, not confined to separate limbs, but affects all equally. It happens, however, that in a slightly motor-apraxic limb, a general uncertainty of ideation is strongly marked. Ideational apraxia is very often associated with agnostic disturbances.

4. *The Predominance of the Left Hemisphere in Controlling Muscular Actions and the Localization of Apractic Disorders*

Heretofore, it has been assumed that the plan of movement rises in the entire brain, that the visual images in both hemispheres and the kinetic centers for all the limbs share it equally, and that this plan is transmitted to the center of the acting limb. This, to a certain extent, is fiction. It has been proved that the special kinetic function of the left arm center and its connection with the rest of the perception centers has not only a peculiar importance for the movement of the right arm, but also for movement in other parts of the body and most surely for those of the left arm. Probably the arm center in the left hemisphere is not only an intermediate station for the stimulation of the arm center in the right hemisphere in purposive movements, but the kinetic memories of the arm center in the left hemisphere are an almost indispensable prop for the *ideational* plan and for movements of the left hand.

As a matter of fact, we find, that in many lesions of the left hemisphere, that paralyze the right hand or make it apraxic, the praxia of the left hand is also affected, whence it is clear, that the direction of both halves of the body in purposive movements is assumed, to a considerable degree, by the left hemisphere.

The left hemisphere predominates, then, as in speech, so also in action as well, even if not in similar degree. Focal injuries that strike,

1. The hand center in the left hemisphere, or

2. The underlying white matter, or

3. The connection of the hand center with other parts of the brain, especially, the temporal, parietal and occipital lobes, cause in the majority of people, besides the effect on the right side of the body (in 1 and 2 mostly paralysis, in 3 apraxia of the right arm) the following dyspraxic symptoms of slighter degrees of severity, in the *left*, unparalyzed, upper extremity:

1. Movements from memory can be performed only in a curtailed fashion, or not at all: the patient can not longer differentiate how one catches a fly, turns a hand-organ, or swings a cane, etc. Especially he can not perform correctly *expressive movements*, as threatening, winking, giving a military salute, throwing kisses, making a long face, etc. More or less distorted movements result, and many *perseveration* miss-reactions occur. If one can ascribe all this to mere amnesia (difficuly in rousing the memory of the meaning and the word), it appears

2. That the patient can not *imitate* movements, so that not only the spontaneous evoking of movement memories is assigned to the left hemisphere, but a direction of the movements of the left hand by the left hemisphere must be maintained.

3. That a few patients manipulate objects wrongly, even with the left

hand. The majority, on the other hand, can when they see and feel the objects, by the help of optic-tactile-kinesthetic signals coming from the object, perform movements, that they could not accomplish from memory alone.

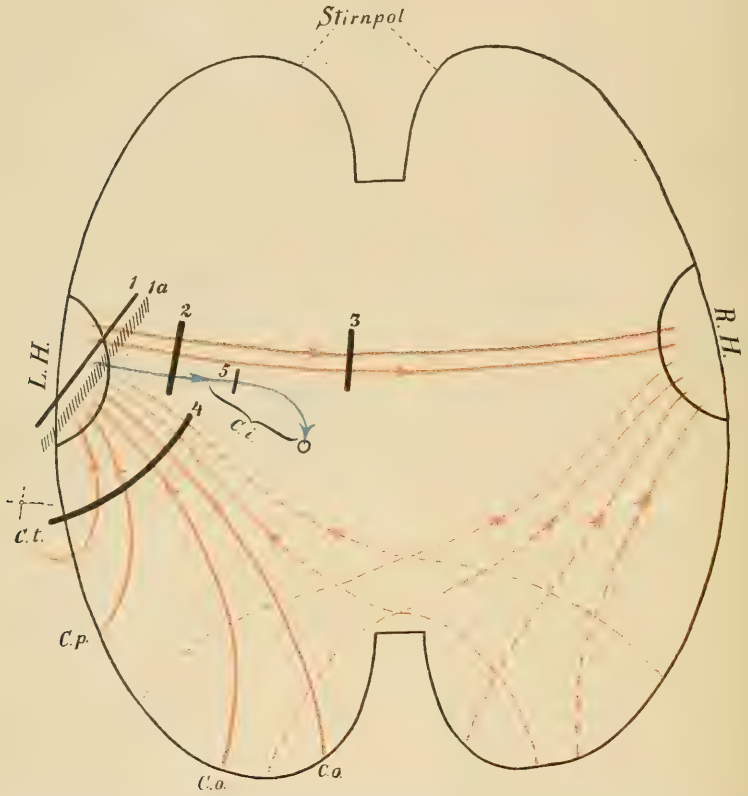


FIG. 156.—Horizontal scheme of the apraxic disturbances. *L.H.*, left brain center of the right hand. *R.H.*, right brain center of the left hand. *C.o.*, *C.p.*, *C.t.*, cortical origin of the occipital, parietal and temporal association fibres to the left brain hand center. The corresponding association paths to the hand center in the right brain as well as those coursing from the right himisphere to the left in red dashes, to point out their subordinate importance. The corpus callosum connections between *L.H.* and *R.H.* are marked by two full-drawn red lines. The blue line which at the end of the arrow deviates from the plane of the figure, represents the projection fibres of *L.H.* The pathway for purposive movements of the right hand is from *C.o.*, *C.p.*, *C.t.*, over *L.H.* through the blue line into the anterior horn cells of the cervical cord. For purposive movements of the left hand principally from *C.o.*, *C.p.*, *C.t.*, over *L. H.* through the corpus callosum to *R.H.*; a less important path leads through the red dash lines to *R.H.*

1. The focus, which destroys completely *L.H.*: paralysis of the right and dyspraxia of the left hand.

1*a*. Slighter lesion of *L.H.* which does not lead to paralysis but destroys the mnemic possession of *L.H.* only: kinetic apraxia of the right and dyspraxia of the left hand. 2. Paralysis of the right and dyspraxia of the left hand. 3. (Focus in the corpus callosum) dyspraxia of the left hand. 4. (Focus behind the hand center in the parietal lobe) ideo-kinetic apraxia of the right and dyspraxia of the left hand. Foci in the left hemisphere located farther backwards and diffuse processes frequently cause ideatory apraxia. 5. Focus in the capsula causes paralysis of the right hand without causing dyspraxia of the left hand. Above the cut: Stirnpol = frontal pole.

The weight of evidence and *anatomic findings* indicate that the right hemisphere in purposive movements of the left arm innervated by it, receives

through the *corpus callosum*, directions from the left hemisphere. *Extensive lesions in the corpus callosum have in a series of cases made the left hand dyspraxic.*

One may suppose, then, that memories of well-learned activities, the free plan of movements, and, finally the supervision of their execution are predominantly the affairs of the left hemisphere and through the corpus callosum are transmitted to the right.

The fact that apraxia of the left hand in focal injuries of the left hemisphere is usually not of a very high degree, and especially often spares the manipulation of objects, proves that the right hemisphere is not *wholly* dependent on the left for praxia, but has a certain measure of kinetic power peculiar to itself, as well as connections with the entire brain.

Eupraxia, then, is dependent on the soundness of a great apparatus, the main part of which is the arm center in the left hemisphere. If this is wholly destroyed (Fig. 156), paralysis of the right arm and dyspraxia of the left appear. If it is but slightly injured (1a) limb-kinetic apraxia of the right arm and again dyspraxia of the left appear. If the white matter underneath the hand center be injured (2), the right arm is again paralyzed, and the left, because the corpus callosum fibres are broken in the white matter, is dyspraxic. If in the parietal lobes, by an extensive focal injury, the connections of the arm center with the temporal parietal, and occipital lobes and the right hemisphere are interrupted, (4) ideo-kinetic apraxia of the right arm appears, associated with slight dyspraxia of the left.

Focal injuries still farther back, in the hindmost part of the temporal, and foremost part of the occipital lobes, as well as diffuse injuries of the brain show ideational apraxia.

If only the corpus callosum is interrupted to a very considerable extent, (3) the left hand is dyspraxic, the right, is neither paralyzed nor dyspraxic. How focal injuries in the *right* hemisphere by robbing the arm center in the right hemisphere of the nervous impulses, that carry the directions (and eventually by destroying the memory), influence praxia of the left hand, has not been definitely ascertained.

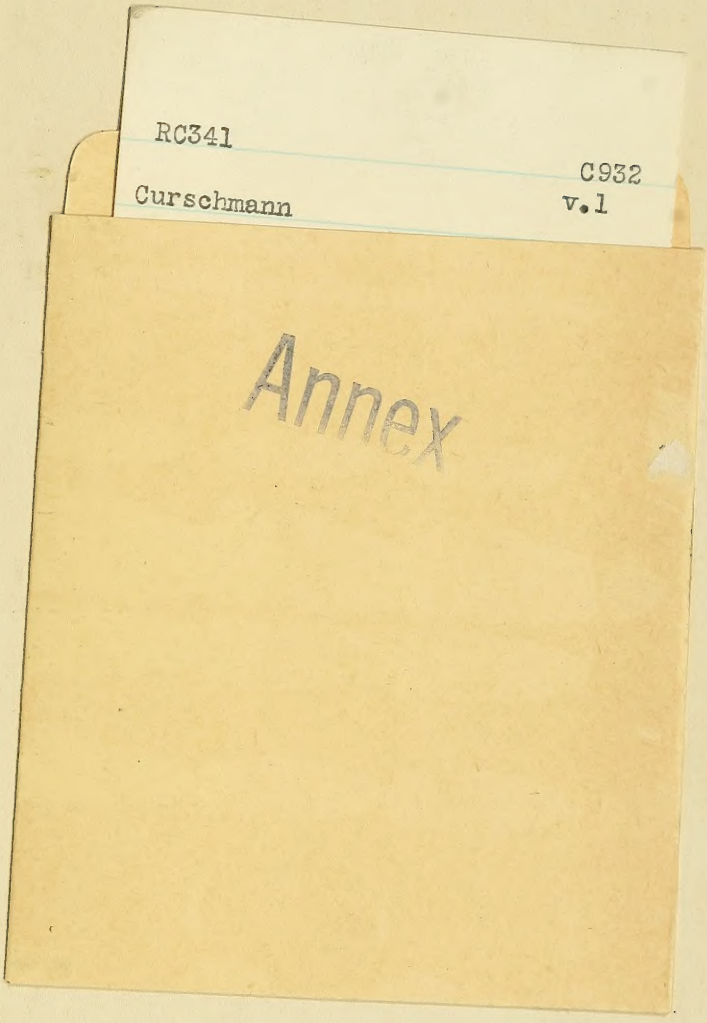
Focal injuries that destroy the *projection fibres* from the inner capsule, and farther down (5), cause no apraxia of the hand on that side, because the corpus callosal fibres are not affected. So that only *supracapsular* focal injuries lead to apraxia. Hence apraxia is a characteristic that *permits us to distinguish* cortical injuries and those in the white matter, from those that are more deeply laid, in the capsules, peduncles, pons, and bulbar region.

A phenomenon has been described under the name mind palsy which stands between actual paralysis and apraxia: a limb can not be moved at will, or only with great difficulty, but shows, through movements, that are performed under peculiar circumstances that it is not really paralyzed, and *by their accuracy,* that it is not apraxic. *It is only a matter of great difficulty in*

innervation. Since the words "mind palsy" (Seelenlaehmung) are used in very different senses, one should call this phenomenon "volition palsy."

Whether, and to what extent, the left *frontal* brain lobe, participates in praxia, is as yet uncertain.

The *left* parietal lobe is of the greatest importance for apraxia. Extensive focal injuries in the left hand center, itself (central convolutions), and in its white matter, *paralyze* mostly the right hand (la is a rare case); apraxia is concealed in the right, and shows itself only in quantitatively slighter disturbances of the left hand. The lesion of the parietal lobe, on the contrary, causes apraxia of the right hand to appear clearly. Lesions of the corpus callosum have considerable influence only as regards the praxia of the *left* hand. So the left parietal lobe is not at all the center for praxia in the sense that there alone purposive movements are "made," but in the sense that it is that place in the brain, in which lesions can interrupt in great number, and with comparative isolation, connective paths most necessary for praxia.



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